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LONG TERM FOLLOW-UP OF MORBIDITY AND QUALITY OF LIFE ASSOCIATED
WITH ISOLATED GASTROSCHISIS

by

Roya Mostafavi, BS

APPROVED:

Claire Singletary, MS, CGC
Supervisory Professor

Syed Shahrukh Hashmi, MD, MPH, PhD

Joan Mastrobattista, MD

Patricia Robbins-Furman, MPH, CGC

KuoJen Tsao, MD

Suzanne Lopez, MD

APPROVED:

Dean, The University of Texas
Graduate School of Biomedical Sciences at Houston

LONG TERM FOLLOW-UP OF MORBIDITY AND QUALITY OF LIFE ASSOCIATED
WITH ISOLATED GASTROSCHISIS

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THESIS

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The University of Texas
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MASTER OF SCIENCE

by

Roya Mostafavi, BS
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LONG TERM FOLLOW-UP OF MORBIDITY AND QUALITY OF LIFE ASSOCIATED WITH ISOLATED GASTROSCHISIS

Publication No. _____

Roya Mostafavi, BS

Supervisory Professor: Claire N. Singletary, MS, CGC

Gastroschisis is a birth defect in which an opening in the abdominal wall allows herniation of the viscera. Prenatal counseling regarding gastroschisis typically discusses that, although these infants often endure a difficult neonatal course, they experience few long-term complications. However, information regarding long-term outcomes is based on limited studies that lack specificity. Therefore, we aimed to study the long-term morbidity and quality of life in children born with gastroschisis in a large and diverse population drawn from the Texas Birth Defects Registry (TBDR). Study packets with informed consent, a questionnaire, and the Pediatric Quality of Life Inventory Generic Core Scale 4.0 (PedsQL 4.0) in English and Spanish were mailed to 1,112 parents of children born with isolated gastroschisis in Texas between 1999 and 2008 via the TBDR. Information was abstracted from the TBDR for 58 mothers of children with gastroschisis who returned study materials. Three hundred fifty five packets were returned to sender, giving a response rate of 7.7%. Children born with gastroschisis had quality of life scores that were not significantly different than expected ($p = 0.981$). However, factors such as having a learning disability ($p = 0.001$) and missing school due to gastrointestinal issues ($p = 0.020$) were found to significantly decrease quality of life. Overall, children with gastroschisis had a significantly increased risk for learning disabilities regardless of whether they were preterm ($p = 0.021$) or full term ($p = 0.021$). Additionally, there appeared to be an increased risk for

auditory impairment in Caucasian children ($p < 0.0005$). Therefore, while overall long-term quality of life is not significantly altered for children born with gastroschisis, the previously unreported increased risk for learning disabilities and possible association with hearing impairment are important findings that should be conveyed to prospective parents.

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Background:

Gastroschisis is a birth defect in which closure of the ventral abdominal wall fails to occur, and the viscera are herniated outward through the opening [1]. The defect is usually to the right of a normal umbilical insertion site [2]. Gastroschisis is thought to occur between the third and fifth weeks of embryonic development. Although the exact cause is unknown, it is most likely due to multifactorial etiology [1, 3].

Prevalence:

The worldwide reported birth prevalence of gastroschisis is currently between 3 in 10, 000 and 5 in 10, 000 live births, and is increasing [4]. In Ireland, the birth prevalence of gastroschisis increased from 1.00 in 10, 000 to 4.94 in 10, 000 live births during the years 1981 to 2000 [5]. Similarly, in South America, the gastroschisis birth prevalence increased from 0.42 in 10, 000 to 5.79 in 10, 000 live births between 1982 and 2006 [5]. A 2010 study conducted by the Texas Birth Defects Registry (TBDR) in collaboration with Texas Tech University found that the statewide birth prevalence of gastroschisis had risen from 1.66 per 10, 000 live births in 1998 to 4.18 per 10, 000 live births between 1999 and 2003 [4]. Additionally, the study found that the prevalence of gastroschisis in 2003 among mothers younger than 20 years of age was 16.14 per 10, 000 live births. Some differences in the prevalence of gastroschisis were reported throughout the geographic regions in Texas as well. After raw data was analyzed, the study reported a higher prevalence of gastroschisis, 4.99 per 10, 000 live births, in babies born to mothers residing along the Texas-Mexico border [4]. Of all of the Texas counties examined, Nueces County had the highest prevalence of gastroschisis. When the health service regions of Texas were examined, the highest prevalence of gastroschisis was observed in the rural Northwest, while the lowest

prevalence was seen in the Houston/Galveston region. After adjusting the data analysis for certain maternal demographic factors, including maternal age, most of the differences in statewide gastroschisis birth prevalence lost statistical significance [4]. The only health region in which a higher prevalence of gastroschisis persisted when compared to the Houston/Galveston referent region was the Austin/Waco region, while the only county that retained a statistically significant higher prevalence of gastroschisis was Nueces County [4]. Also, after adjustment for maternal demographic factors, there were no significant differences in overall prevalence of gastroschisis when comparing the rural and urban areas of the state [4].

Etiology Hypotheses:

Although many hypotheses have been proposed in efforts to explain the origin and embryological cause of gastroschisis, the definitive cause of the birth defect remains unknown. One of the hypotheses concerning the pathogenesis of gastroschisis proposes that the defect occurs as result of a teratogenic exposure that interferes with mesenchymal differentiation during the fourth post-conceptual week [1]. The hypothesis of failed mesenchymal differentiation was succeeded by a hypothesis that focused on rupture of the umbilical ring as a cause of gastroschisis. This theory suggests that the amniotic membrane of the umbilical ring ruptures at some point in fetal development, leading to the exposure of viscera to the outside environment [1]. Other potential etiologies of gastroschisis focus on vascular mechanisms that may be responsible for the abdominal wall defect. One of these vascular hypotheses suggests that twisting or coiling of the right umbilical vein may occur, inhibiting growth of the mesenchyme and formation of skin in the region around the

umbilicus. The resulting defect would then allow the abdominal contents to be exposed to the extra-abdominal cavity [1]. Another etiological theory for gastroschisis proposes that vascular compromise may be the cause of the defect; however, this theory suggests that gastroschisis is the result of a disruption of the vitelline artery. The disruption is thought to allow for necrosis at the base of the umbilicus and resultant visceral herniation [1]. Another possible etiology of gastroschisis is failure of the yolk sac and associated vitelline structures to be incorporated into the umbilical cord during normal development. Isolation of the vitelline structures outside of the umbilicus is thought to prevent the gut from herniating into the umbilicus, as is expected in normal development [6]. One other hypothesis for the pathogenesis of gastroschisis advocates that the defect is a result of incomplete folding of the body wall, allowing for herniation of intestinal loops into the amniotic cavity [1]. Further research is needed to determine if any of the proposed hypotheses can definitively explain the etiology of gastroschisis.

Risk Factors:

Various risk factors have been investigated in relation to gastroschisis. The most consistently identified risk factor for gastroschisis is young maternal age, which confers a significantly higher risk of having a child with gastroschisis [3, 5, 7]. A 2004 study revealed that women who are between the ages of 14 to 19 years are at a seven times higher risk of having a child with gastroschisis when compared to women 25 to 29 years old. Young paternal age has also been associated with an increased risk for gastroschisis; however, this association is not as strong or as well-studied as the association between young maternal age and the occurrence of gastroschisis [3]. Increased maternal BMI correlates with a decreased

risk of gastroschisis. A 2007 study that adjusted for maternal age found that maternal obesity is correlated with an adjusted odds ratio of 0.19 (95% CI, 0.10-0.34) for having a child born with gastroschisis, revealing a significantly lower risk for infants of these mothers to have gastroschisis [8]. However, the age-adjusted odds ratio for mothers with BMIs below 18.5 to have a child with gastroschisis was not statistically significant (OR: 0.85; 95% CI, 0.58-1.23), indicating that low maternal BMI is not a significant risk factor for gastroschisis [8].

Other proposed risk factors for gastroschisis include low socioeconomic status, maternal genitourinary tract infections immediately before pregnancy or during the first trimester, maternal x-ray exposure early in pregnancy, maternal exposure to solvents, maternal aspirin, ibuprofen, and acetaminophen use, maternal cigarette smoking, maternal alcohol consumption, maternal use of illicit drugs, particularly vasoconstrictive drugs; and maternal use of decongestants [3]. Certain maternal dietary factors are correlated with an increased risk for having a child with gastroschisis; particularly, low levels of glutathione and α -carotene, which are antioxidants found in many fruits and vegetables; low protein; low zinc; and high nitrosamines, which are found in cured meats and beer [3]. High levels of maternal fat intake may be a risk factor for gastroschisis, but a clear association has not been established [3]. It is also unclear whether oral contraceptive use at the time of conception or early in the first trimester is a risk factor for having a child with gastroschisis. Short periods of cohabitation between partners and multiple changes in paternity between pregnancies have been suggested to correlate with higher risks for gastroschisis in a few studies; however, data is inconclusive and may be confounded by lifestyle factors [3]. These factors may be associated with immune maladaptation and a modified maternal

immune response towards pregnancy due to the amount of time that the mother is exposed to paternal antigens [3, 9]. Additionally, based on observed familial cases of gastroschisis, the recurrence risk of gastroschisis for siblings is 3.5%, suggesting a genetic component for the defect [3, 8].

It is unknown whether certain ethnic groups are at an increased risk to have a child with gastroschisis [3]. In some studies, the risk of gastroschisis has been associated with maternal ethnicity with the highest risk being in Hispanics and lowest in African Americans. However, upon adjustment for other known risk factors, the observed differences between ethnicities dissolve [3, 10].

Prenatal Diagnosis and Management:

Gastroschisis is a birth defect that is diagnosed by prenatal ultrasound, with 83% to 96.5% of cases detected before the third trimester of pregnancy [11, 12]. Gastroschisis may be suspected if maternal serum screening during the second trimester of pregnancy reveals an elevated maternal serum alpha-fetoprotein (AFP), [13]. Routine anatomy scans scheduled in the second trimester of pregnancy may also detect many cases of prenatally diagnosed gastroschisis. Gastroschisis is not commonly diagnosed with certainty before 14 weeks gestational age [14]. In gastroschisis, free-floating small intestine is visible by ultrasound, although the stomach and large intestine may also appear outside of the abdominal cavity in some cases. Rarely, the liver may be involved in the gastroschisis [14, 15]. The increased risks in pregnancies involving gastroschisis include preterm labor, intrauterine growth restriction, abnormal levels of amniotic fluid, and an increased risk of stillbirth [11, 14]. Given these increased risks, once the diagnosis of gastroschisis is made, it

is generally recommended that the pregnancy be managed by a team of obstetric providers, maternal-fetal medicine specialists, pediatric surgeons, neonatologists, and genetic counselors at a tertiary care center [14]. To evaluate for possible complications, pregnancies involving gastroschisis are usually monitored with regular non-stress tests (NSTs) and biophysical profiles (BPP) beginning near 30 weeks gestation, in addition to serial fetal growth ultrasounds [13, 14]. If threatened by preterm birth between 24 and 34 weeks, antenatal corticosteroids may be administered to enhance fetal lung maturity [14].

Multiple studies have attempted to identify prenatal markers that can be used to detect the severity of gastroschisis. Markers examined include intra-abdominal bowel dilation, extra-abdominal bowel dilation, whether or not the fetus was small for gestational age, thickening of intestinal walls, stomach dilation, hyperperistalsis, oligohydramnios, and polyhydramnios [16-18]. Of the markers examined, intra-abdominal bowel dilation was the only consistent statistically significant marker of postnatal complications [16, 18]. In one study, intrauterine growth restriction was found to be an indicator of adverse outcomes in pregnancies complicated by gastroschisis [17].

The effect of prenatal diagnosis of gastroschisis on outcome of live-born infants has been examined. Studies have shown that live-born babies with gastroschisis diagnosed prenatally and those diagnosed at birth had no differences in postnatal outcomes, such as time to first enteral feeds, time to abdominal closure, days on antibiotics, incidence of necrotizing enterocolitis, and length of hospital stay [11, 19]. However, the complications related to preterm delivery and bowel atresia were found to affect the postnatal outcomes in infants with gastroschisis [11]. Therefore, Snyder et al. [19] stated that infants with prenatally diagnosed gastroschisis were more likely to be delivered at term or at later

gestational ages. Also, prenatal diagnosis of gastroschisis may contribute to lower rates of intrauterine death and increased detection of intestinal abnormalities [11]. Currently, prenatal counseling regarding gastroschisis takes an optimistic approach and proposes that, although these infants often endure prolonged hospital stays and increased dependence on parenteral feeding in the neonatal period, they experience few long-term complications related to gastroschisis. However, information regarding the long-term outcomes of gastroschisis is based on limited studies that lack specificity [14].

Surgical Correction:

Once a baby with gastroschisis is delivered, the defect must be surgically closed. Primary closure of the abdominal wall defect usually requires only one operation, while staged reduction involves the use of a silo and at least two surgical operations [20]. When possible, primary closure is the preferred method of repairing gastroschisis [21]. Primary closure can be subdivided into both primary fascial closure and primary sutureless closure [20]. Sutureless closure differs from fascial closure in that it involves covering the defect with the remaining umbilicus and a synthetic dressing that holds it in place, while fascial closure involves placement of sutures to hold the umbilicus in place. The sutureless form of primary closure was introduced in 2004 [20]. When the defect is complicated by excessive inflammation and primary closure cannot be performed, staged reduction is the method of closure [22]. Staged reduction can be carried out using either a customized silo or the more recent preformed silo, introduced in 1995 [20, 22]. A 2006 study in Texas reported that infants who required staged reduction for correction of their gastroschisis experienced significantly more gastrointestinal complications than those who underwent primary closure.

The same study reported that staged reduction was also associated with longer times on parenteral nutrition and a later start to full feeds [21]. Other studies have also shown staged reduction to be significantly associated with a longer duration of postoperative metabolic acidosis and positive fluid balance [20]. Infants who undergo staged reduction are also more likely to spend more time on mechanical ventilation, have a longer length of hospital stay, and incur greater costs in hospital charges than infants in whom primary closure was attempted [23].

A recent study conducted by South, Wessel, Sberna, Patel, & Morrow [24] at Cincinnati Children's Hospital described the complications experienced by children born with gastroschisis between 2006 and 2008. The study focused on the frequency of hospital readmission for children born with gastroschisis. In the South et al. [24] study, infants who underwent primary closure were more likely to be readmitted to the hospital for bowel obstruction after initial discharge. The authors suggested that further examination of the relationship between the method of gastroschisis repair and long-term morbidities is necessary to clarify the expected prognosis for these children [24].

Neonatal Outcome:

Currently, the overall survival rate for gastroschisis is greater than 90%, with about 5-10% of affected pregnancies resulting in fetal deaths and approximately 5% of babies born with gastroschisis dying in the first few months of life [7, 13]. The TBDR reports that between 1999 and 2007, 92% of gastroschisis cases were liveborn, 6% were stillborn, and less than 4% underwent pregnancy termination [10]. Studies assessing cases of gastroschisis in the neonatal period have identified factors that do not affect immediate outcome. The

mode of delivery has not been shown to be a significant factor in the neonatal outcome of infants with gastroschisis [19]. Multiple studies have shown that caesarian deliveries do not confer significant advantages over vaginal delivery of infants with gastroschisis [11, 19, 25, 26]. Also, delivery of fetuses with gastroschisis prior to and after 36 weeks gestation does not significantly affect intestinal injury [27]. Recent studies report no differences in time to closure when comparing gestational ages of these infants at delivery, although older gestational age at closure is a significant predictor of early enteral feeding [19]. The effect of location of delivery on the outcome of fetuses with gastroschisis has not recently been evaluated. Studies in the early 1990's report that fetuses with gastroschisis born in regional care facilities experience comparable neonatal outcomes to babies who were transferred to these centers after birth [28, 29]. Both Nicholls et al. [29] and Stoodley et al. [28] did not find significant differences in the gestational age at delivery, birth weights, caesarian delivery rates, time to operation, mortality rates, rates of primary closure, or times to full enteral feeding when comparing infants born in tertiary care obstetric centers to infants born in community hospitals and subsequently transferred to tertiary centers. However, both studies reported significantly higher rates of prenatal diagnosis of gastroschisis occurring in the infants delivered at tertiary centers when compared to the peripheral group.

Long-term Outcome Studies of Gastroschisis:

Multiple studies have examined the neonatal outcome of infants with gastroschisis, but very few have investigated long-term morbidity and quality of life in these children. Most of the recent studies assessing quality of life in children with gastroschisis have focused on European populations [30, 31]. In addition, these studies assessed quality of life

and morbidity related to both gastroschisis and omphalocele, another birth defect involving the abdominal wall [30, 31]. Koivusalo et al. [31] focused solely on a study population of Finnish individuals, of which only 11 had gastroschisis. In this population, acquired diseases, which included cholelithiasis, asthma, gastroesophageal reflux, and food allergies, were seen to occur in the gastroschisis and omphalocele population at a similar frequency as in the general population [31]. Eighty-eight percent of ascertained individuals with gastroschisis or omphalocele considered themselves to be in good health, while 12% of the individuals with gastroschisis or omphalocele were found to have low self-esteem [31]. The most common sources of morbidity in the study population were related to the surgical scar and to functional gastrointestinal disorders, while quality of life and education level in the study population did not differ from that of the general population [31].

A 2008 study by Henrich et al. examined 22 children born with gastroschisis for long-term follow up (mean duration of time to follow up was 6.3 years) and found that 7.1% of the children followed had recurrent gastrointestinal issues. Two children reported difficulty with physical activity [30]. Eighteen of twenty-two (82%) of the children in this study reported the cosmetic results of the scar to be “good” [30]. Seven of twenty-two (32%) had delay in their speech and walking [30]. The study also found that 9% of these children had weights below the third percentile and 14% had heights below the third percentile [30]. All delays in development observed in these children resolved in the first two years of life [30].

The majority of American studies describing long-term quality of life in children born with gastroschisis evaluated small sample populations consisting of both gastroschisis and omphalocele patients born before the advent of new methods of surgical and medical

management [32-34]. These studies concluded that children with repaired gastroschisis experienced a quality of life similar to that of the general population [32]. However, some children with repaired gastroschisis required surgical intervention for gastrointestinal complications, such as intestinal atresia and abdominal wall hernias [32-34]. The absence of an umbilicus after repair was also found to be a concern in the Tunnel et al. [32] and Davies & Stringer [33] studies.

Quality of Life:

Although multiple studies have examined quality of life associated with different medical conditions, no globally accepted definition of “quality of life” exists [35, 36]. Many of the definitions of “quality of life” agree that there are multiple variables that should be incorporated into a quality of life assessment [36]. One definition model of “quality of life” proposed by Flanagan in 1978 divided quality of life into five domains: physical and material well-being, interpersonal relationships, social and civic activities, personal fulfillment and growth, and recreational activities [35].

Health-related quality of life is a term often used when examining the effects of disease or chronic conditions on an individual’s life [36, 37]. As defined by the World Health Organization, “health” is “a state of complete physical, mental, and social well-being, and not merely the absence of disease or infirmity” [35]. Taylor et al. [36] point out that quality of life is impacted differently by various health conditions, because different conditions have varying effects on an individual’s development. Therefore, it is important to evaluate quality of life separately for different conditions.

The lack of long-term follow up information on children with repaired gastroschisis differentiates this condition from other birth defects and genetic conditions in which quality of life issues are much more well-defined. For example, studies focused on long term follow up of isolated cleft lip and palate have shown that these children do not experience significant differences in quality of life as it relates to health or major psychosocial concerns [38, 39]. The assessment of health and health-related concerns for these cleft lip and palate patients is generally within normal limits, although psychosocial issues related to a decreased sense of self-worth during adolescence sometimes occurs [39].

In contrast, quality of life is more significantly affected in individuals with more severe genetic conditions. For example, sickle cell disease is an autosomal recessive genetic disorder where the individual's quality of life is grossly affected [40]. These patients experience a number of health complications, such as chronic pain, avascular necrosis in their joints, acute chest syndrome, leg ulcers, asthma, and hypertension [41]. Patients with sickle cell disease frequently require long-term opioids and antidepressant medications [41]. The life expectancy for individuals with sickle cell disease is greatly reduced in comparison to the general population [40]. Thus, the impact of a genetic condition or birth defect varies widely.

Limitations of Counseling Long-term Outcomes and Qualities of Life Associated with Gastroschisis:

Given the variability in outcomes and quality of life among patients with different genetic conditions and birth defects, both prenatal and postnatal counseling of the affected child's parents must be specific to the particular condition. With so little information

currently available about the long-term implications of gastroschisis, accurate counseling regarding this birth defect remains challenging. The lack of long-term follow up information on a large cohort of children with repaired gastroschisis warrants a descriptive study of the health complications and quality of life experienced beyond the neonatal period. Therefore, our specific aim is to study the long term morbidity and quality of life issues in children born with gastroschisis in a large and diverse population drawn from the TBDR.

Materials and Methods:

Study Participants:

Cases for this study were identified and contacted in collaboration with the TBDR. These cases included liveborn infants with isolated gastroschisis delivered between 1999 and 2008 in Texas, with a British Paediatric Association (BPA) diagnosis code of 756.710 indicating isolated gastroschisis. Cases with a "possible/probable" diagnosis of gastroschisis, those that were stillborn, and non-isolated cases of gastroschisis were excluded from the study.

All identified cases that met inclusion criteria were mailed a letter of invitation (Appendix A), a letter of informed consent (Appendix B), a questionnaire that was designed for this study (Appendix C), and the parent-proxy reports of the Pediatric Quality of Life Inventory Generic Core Scale 4.0 (PedsQL 4.0) for children in the following age groups: 2-4 years old, 5-7 years old, and 8-12 years old (Appendix D). For those who decided to participate in the study, the questionnaire and PedsQL 4.0 scale were filled out by the parent/proxy of the child with gastroschisis and returned by mail.

All study materials were mailed out in both English and Spanish by the TBDR. The questionnaire developed for this study was translated into Spanish by the Texas Department of State Health Services Language Services professionals. All Spanish responses to open-ended questions on the study questionnaires were translated into English by Jennifer Lemons, a bilingual certified genetic counselor at the University of Texas Health Science Center at Houston. All Spanish study documents are available in Appendix E.

Institutional Review Board (IRB) approval for this study was obtained from both the Texas Department of State Health Services (IRB # 11-065) and the University of Texas Health Science Center at Houston (UT IRB HSC-MS-11-0387).

Study Instruments:

Anonymous questionnaires and PedsQL 4.0 scales were used to assess morbidity and quality of life in patients born with gastroschisis. The study packets were mailed in February 2012 to the most recent addresses of cases recorded in the TBDR. The primary caregiver of the child born with gastroschisis was asked to fill out each questionnaire and PedsQL 4.0 as accurately as possible. Each study packet mailed was given a unique identifier corresponding to a case number in the TBDR to protect the privacy of all participants. Therefore, protected health information held by the TBDR was not released to the Principal Investigator (PI) until a completed survey indicating consent to participate was received by the PI. The unique identifier was used to link questionnaire responses to the demographic and medical history information from the TBDR for each participant. Information abstracted from the TBDR included: pregnancy/prenatal history, information regarding delivery, information regarding surgical correction of the gastroschisis, and information regarding infant/fetal complications. The questionnaire contained multiple choice and free-response questions regarding the pregnancy/prenatal history, demographic information about the family with whom the child lives, surgical history of the child, cognitive functioning of the child, and health complications in the child born with gastroschisis. The questionnaire also asked parents to describe what they perceived to be

"good quality of life" and to express how they perceived their child's quality of life. All answers to these open-ended response questions are available in Appendix F.

Quality of life of gastroschisis patients was measured using the parent-proxy reports of the PedsQL 4.0 scale, a standardized assessment tool validated in both English and Spanish-speaking populations. This tool consists of Likert scale questions that are aimed at analyzing physical functioning, emotional functioning, social functioning, and school functioning to determine overall health-related quality of life. The PedsQL 4.0 incorporates measures of physical, mental, and social health, thereby fulfilling the requirements set forth by the World Health Organization (WHO) in 1948 for a pediatric health-related quality of life measurement instrument [42]. A 2007 study by Varni et al. assessed reliability and validity of the PedsQL 4.0 parent-proxy reports, and found that the majority of the scales in the PedsQL 4.0 showed α coefficients above the minimum reliability standard of 0.70 that is required for making group comparisons [43]. The PedsQL 4.0 scales for children five to twelve years old contain 23 Likert scale questions, while the PedsQL 4.0 scale for children two to four years old contains 21 Likert scale questions.

Statistical Analysis:

The PedsQL 4.0 forms were scored according to the standard PedsQL 4.0 scoring manual. The 0-4 point scale items from the PedsQL 4.0 questionnaire were transformed to a 0 (lowest quality of life) to 100 (highest quality of life) point scale as follows: 0=100, 1=75, 2=50, 3=25, and 4=0 (pedsq.org). Means were calculated for each Scale Score by dividing the sum of the items by the number of items answered in order to account for missing data fields. However, Scale scores were not calculated for any scale in which 50%

or more of the data fields in the scale were left blank. In cases where missing fields existed, but did not exceed 50% of the scale, the mean for the Scale Score was calculated by summing the scores for all items in the scale, and then dividing by the number of items in the scale minus the number of missing fields. The means of Psychosocial Health Summary Scores were computed by summing items in the Emotional, Social, and School Functioning scales and then dividing by the number of items answered in these scales. We measured physical health by computing the mean for the Physical Functioning Scale Score. Total Scale Scores were calculated by summing the items in all scales and then dividing by the number of items answered [42].

All collected data from the questionnaire and the TBDR was entered into a Microsoft Excel spreadsheet. Descriptive statistics including means, standard deviations, medians, and interquartile ranges (IQR) for demographic variables and survey responses were calculated using STATA version 10.0 (STATA Corp., College Station, TX). Wilcoxon rank sum tests and Kruskal-Wallis tests were used to determine whether or not PedsQL 4.0 quality of life scores were significantly influenced by demographic variables or past medical history. For morbidities reported frequently in our population binomial tests and one-sample t-tests were used to determine whether the prevalences of these morbidities in our gastroschisis patients were significantly higher than prevalences reported in the general population. One-sample t-tests were also used to determine whether the quality of life scores in the gastroschisis population differed significantly from expected scores. One sample tests of proportion were used to determine whether or not significant differences in maternal age, maternal ethnicity, and infant gender existed between our population and the population of gastroschisis cases in the TBDR. Chi-square tests were used to determine

whether the frequencies of select perinatal complications were significantly different between gestational age groups. A p-value of less than 0.05 was considered significant for all statistical analyses.

Results:

A total of 1,112 study packets were mailed out to parents of children born in Texas between 1999 and 2008 with isolated gastroschisis. Data collection for this study began on February 7, 2012 and ended on March 20, 2012. During this time, fifty-eight responses were received by the Principal Investigator. Approximately 355 envelopes were returned to the TBDR mailroom due to invalid addresses. Therefore, this study had a response rate of 7.7%.

Once a questionnaire and/or PedsQL 4.0 scale was returned by a participant, data for that participant, including demographic, prenatal, and postnatal information, was requested from the TBDR. Data from the questionnaires, PedsQL 4.0 scales, and the TBDR were analyzed to describe the quality of life and common morbidities experienced by individuals born with gastroschisis and to determine whether any predictors of their quality of life or frequent morbidities exist.

Demographics:

A total of fifty-eight responses were analyzed for this study. Fifty (86.21%) were returned in English, and eight (13.79%) were received in Spanish. Of the fifty-eight respondents, all identified themselves as the mothers of the children with gastroschisis. Demographic information for these mothers is summarized in Table 1. The majority of mothers (74.14%) were twenty-four years old or younger when they delivered their child with gastroschisis. Calculation of pre-pregnancy BMIs of the mothers was also performed, revealing that most mothers (67.24%) had pre-pregnancy BMIs within the normal range of 18.5-24.9. Twenty-eight (48.28%) mothers were of white non-Hispanic ethnicity, four (6.9%) mothers were of black non-Hispanic ethnicity, and twenty-six (44.83%) mothers

were of Hispanic ethnicity. Maternal education levels were obtained from the TBDR.

Sixteen mothers (27.59%) had less than a high school education, twenty mothers (34.48%) had completed high school, and twenty-two (37.93%) mothers had more than a high school education. Maternal ages and ethnicities, as well as infant genders, in this study population were not significantly different from those in the overall TBDR gastroschisis population (Table 2) [44].

Table 1: Maternal Demographic Information

Maternal Demographics					
Maternal Age at Delivery		Pre-Pregnancy BMI		Maternal Ethnicity	
Age	n (%)	BMI	n (%)	Ethnicity	n (%)
<20	19 (32.76%)	<18.5	2 (3.45%)	Non-Hispanic White	28 (48.28%)
21-24	24 (41.38%)	18.5-24.9	39 (67.24%)	African American	4 (6.9%)
25-29	11 (18.97%)	25.0-29.9	8 (13.79%)	Hispanic	26 (44.83%)
30-35	4 (6.9%)	≥30.0	7(12.07%)		
		Missing	2 (3.45%)		
Maternal Education		Annual Household Income		Language (based on survey response)	
Education Level	n (%)	Income	n (%)	Language	n (%)
Less Than High School	16 (27.59%)	<25,000	15 (25.86%)	English	50 (86.21%)
High School	20 (34.48%)	25,000-49,999	16 (27.59%)		
Greater Than High School	22 (37.93%)	50,000-74,999	10 (17.24%)	Spanish	8 (13.79%)
		>75,000	12 (20.69%)		
		Missing	5 (8.62%)		

Table 2: Lack of Significant Difference between the Study Population and TBDR Population

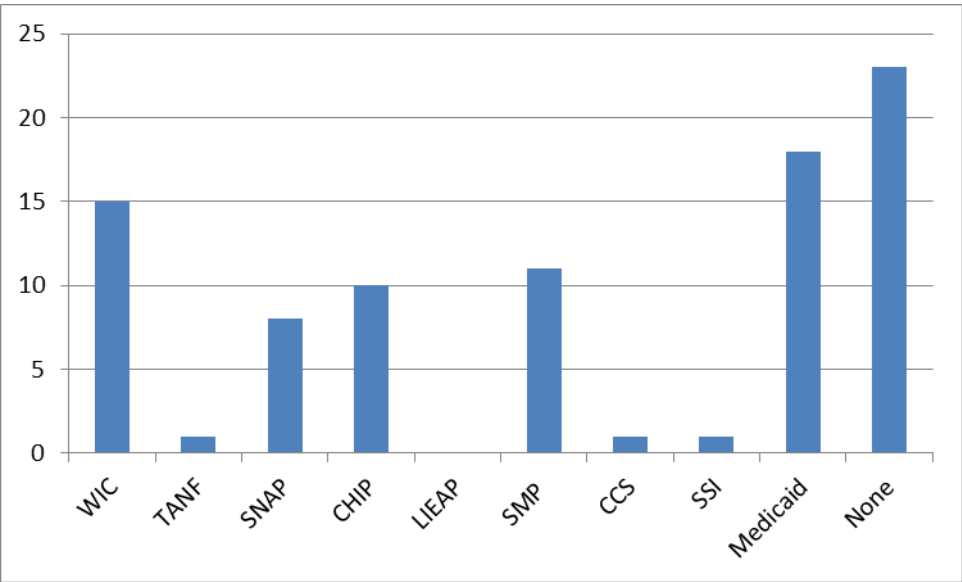
Study Population Variation from the Texas Birth Defects Registry					
Maternal Age at Delivery		Maternal Ethnicity		Infant Gender	
Age	p-value	Ethnicity	p-value	Gender	p-value
<20	0.807	Non-Hispanic White	0.086	Male	0.142
21-24	0.449	African American	0.487	Female	0.783
25-29	0.231	Hispanic	0.868		
≥30	0.451				

Socioeconomic Status and Access to Resources

Mothers were asked to report annual household income on the questionnaire. Fifteen (25.86%) mothers reported an annual household income of less than \$25,000, sixteen (27.59%) mothers reported an annual household income of \$25,000 to \$49,999, ten (17.24%) mothers reported an annual household income of \$50,000 to \$74,999, and twelve (20.69%) mothers reported an annual household income of greater than \$75,000. Five (8.62%) mothers did not report annual household income. To further clarify financial status and access to resources, mothers were asked whether or not they received any of the following sources of financial aid: Women, Infants, and Children vouchers (WIC), (n = 15), Temporary Assistance to Needy Families (TANF), (n = 1), Supplemental Nutrition Assistance Program (SNAP), (n = 8), Children's Health Insurance Program (CHIP), (n = 10), School Meals Program (SMP), (n = 11), Low Income Energy Assistance Program

(LIEAP), (n = 0), Child Care Subsidies (CCS), (n = 1), Supplemental Security Income (SSI), (n = 1), or Medicaid (n = 18). The number of women receiving each source of financial aid is represented in Figure 1. Since respondents could select more than one source of financial aid, these categories are not mutually exclusive. Twenty-three mothers reported receiving no source of financial aid.

Figure 1: Financial Aid Sources Received by Mothers of Gastroschisis Cases



Prenatal and Birth History:

Prenatal and birth history for the gastroschisis cases was obtained from the TBDR. Thirty-six (62.07%) gastroschisis cases were born to nulliparous women, fifteen (25.86%) were born to women with one previous pregnancy, three (5.17%) were born to women with two previous pregnancies, two (3.45%) were born to women with three previous pregnancies, and two (3.45%) gastroschisis cases were born to women with four previous pregnancies. The average gestational age (by clinical exam) at birth for our gastroschisis population was 36 weeks (SD 1.68). On the questionnaire, twenty (34.48%) mothers reported that their children were born full-term (≥ 37 weeks), thirty-seven (63.79%) mothers reported that their children were born premature (before 37 weeks), and one mother (1.72%) did not answer this question. The rate of prematurity (63.79%) in our study population is consistent with literature reporting that approximately two-thirds of infants with gastroschisis are born premature [24, 45]. Mothers were also asked to indicate route of delivery on the questionnaire. Twenty-two (37.93%) mothers reported vaginal delivery, thirty-five (60.34%) reported delivery by Caesarian section, and one mother (1.72%) did not specify route of delivery. Likewise, mothers were asked to indicate whether delivery was spontaneous, planned, or induced for medical reasons. An “other” option was also given so that mothers could write in the delivery plan for their child if necessary. The number of mothers who reported delivery as either spontaneous or planned were approximately equal, nineteen (32.76%) and twenty (34.48%) respectively. Thirteen (22.41%) mothers indicated that they were induced for medical reasons. One mother indicated that she did not know the delivery plan for her child. Three (5.17%) mothers selected the “other” option and two (3.45%) mothers did not answer the question. Of note, some mothers did not select the

"other" option, but did write indications for their induced deliveries. Mothers who selected the "other" option or included comments stated that they were induced because of preeclampsia, reduced fetal movement, restricted intestinal blood flow, or because the cord was wrapped around the baby's neck. One mother was admitted for low fluid and had contractions the next day, and one mother indicated that her water broke.

Forty (68.96%) of the gastroschisis cases had an Apgar score of 8 or 9 at one minute after birth, while fifty-one (87.94%) of the gastroschisis cases had an Apgar score of 8 or 9 at five minutes after birth. Twenty-three (39.66%) returned questionnaires were from families in which the child with gastroschisis was female and thirty-five (60.34%) in which the child was male. Some of the cases in this study experienced fetal or infant complications that were recorded by the TBDR. Fetal and infant complications observed in this study population are presented in Table 3. The frequencies of complications in the gastroschisis patients are shown in Figure 2. Of the complications that occurred in seven or more patients, anemia ($p < 0.0005$), hyperbilirubinemia ($p = 0.041$), and gastroesophageal reflux ($p = 0.045$) were significantly more common in preterm infants.

Figure 2: Number of Fetal/Infant Complications Experienced by Gastroschisis Cases

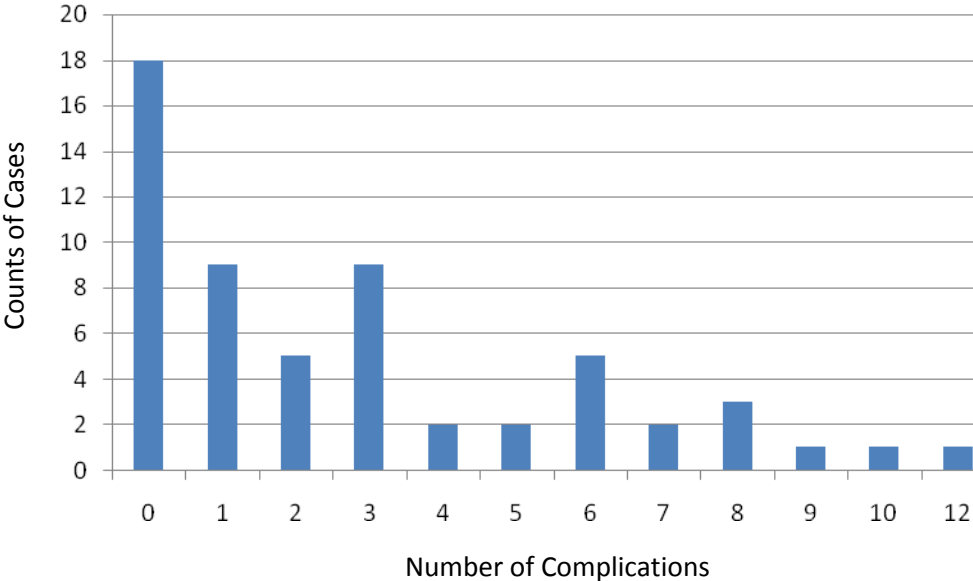


Table 3: Frequency of Reported Infant and Fetal Complications

Complication	# of Patients
Anemia	16
Intubation	15
Respiratory Distress Syndrome	11
Jaundice	10
Hyperbilirubinemia	9
Meconium Other	9
Sepsis	9
Gastroesophageal Reflux	8
Meconium NOS	8
Feeding Problems	7
Oligohydramnios	6
Intrauterine Growth Restriction	5
Apnea	4
Thrombocytopenia	4
Hyaline Membrane Disease	3
Umbilical Cord: wrapped around body part	3
Breech Presentation (any type)	2
Fetal Bradycardia	2
Fever	2
Hypoglycemia	2
Hypotension	2
Hypovolemia	2
Intraventricular Hemorrhage	2
Polyhydramnios	2
Poor Feeder	2
Respiratory Depression	2
Decels	1
Decreased Fetal Movement	1
Dehydration	1
Fetal Tachycardia	1
Hypoperfusion	1
Meconium Aspiration	1
Metabolic Acidosis	1
Necrotizing Enterocolitis	1
Pneumonia	1
Poor Muscle Tone	1
Poor Suck	1
Persistent Pulmonary Hypertension of the Newborn	1
Respiratory Failure	1
Retinopathy of Prematurity	1
Shock	1
Urinary Tract Infection	1

The gastroschisis cases in this study population were born throughout the state of Texas as indicated by information in the TBDR. In regards to the gastroschisis cases included in this study, the number of cases born in each of the eleven health service regions of Texas is shown below in Figure 3. Table 4 indicates the total number of infants with gastroschisis delivered in each health service region between 1999 and 2008.

Figure 3: Number of Cases Born in Each Health Service Region

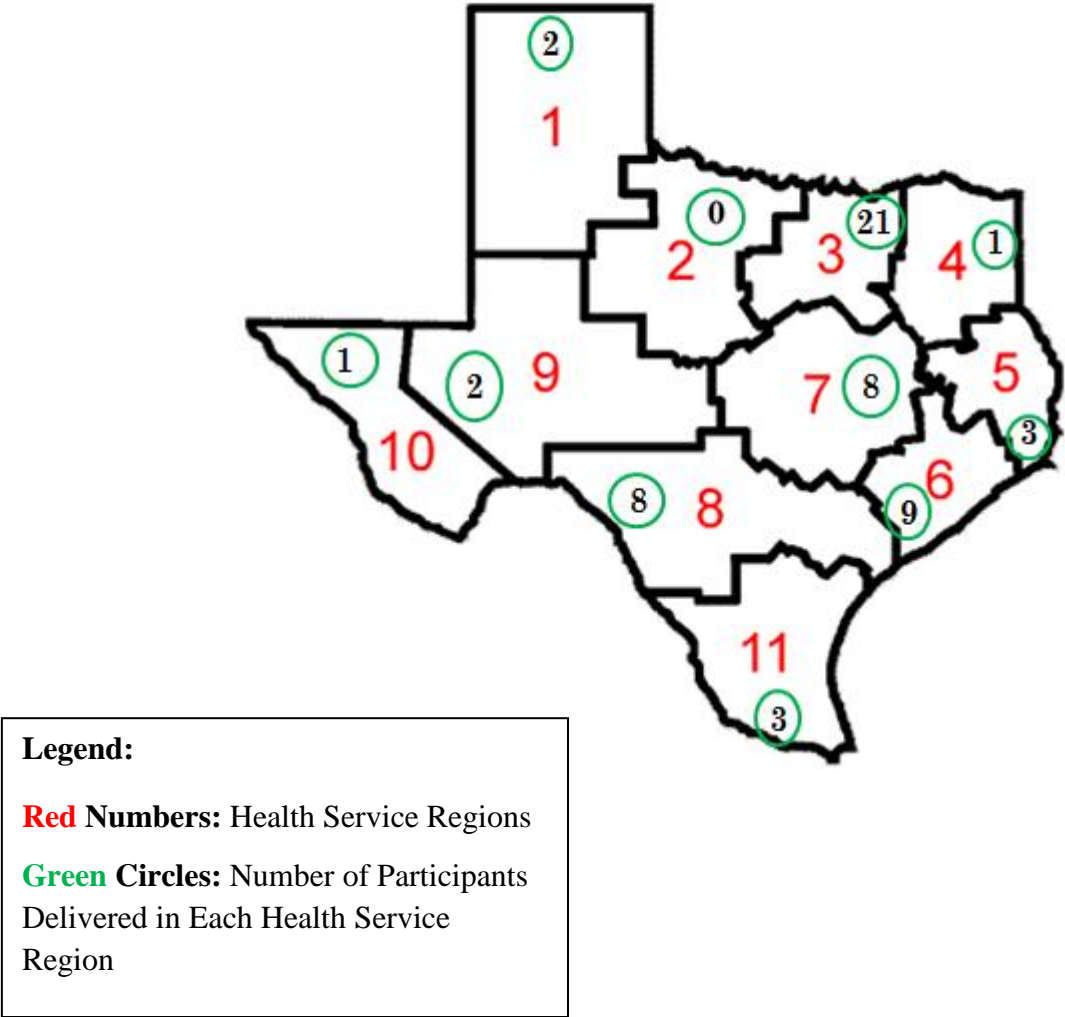


Table 4: Total Number of Gastroschisis Deliveries in Texas Health Service Regions between 1999 and 2008

Health Service Region	Number of Gastroschisis Cases Delivered between 1999 and 2008*
Health Service Region 1	72
Health Service Region 2	51
Health Service Region 3	472
Health Service Region 4	64
Health Service Region 5	55
Health Service Region 6	324
Health Service Region 7	199
Health Service Region 8	228
Health Service Region 9	60
Health Service Region 10	78
Health Service Region 11	231

*Includes ALL cases of gastroschisis delivered between 1999 and 2008. Not all of these cases met inclusion criteria for this study.

Postnatal Medical History of the Gastroschisis Cases:

Medical and surgical histories of our gastroschisis cases were ascertained through the questionnaire. Forty-four (75.86%) mothers reported that their children received postnatal care in the same hospital in which they were delivered, while twelve (20.69%) mothers indicated that their children were transferred to different hospitals after birth. One (1.72%) stated that her child was cared for in the same hospital in which delivery occurred, but was also transferred to a different hospital for care.

On the questionnaire, mothers were also asked to indicate the types of surgeries performed to repair the gastroschises in their children. Twenty-nine (50%) of our study

population cases underwent primary closure. Twenty-six (44.83%) of the gastroschisis cases were repaired over a few days with the use of a silo, while two (3.45%) of the cases were repaired over the course of several weeks using a silo. One mother did not indicate the type of surgery used to repair her child's gastroschisis.

The questionnaire developed for this study also asked mothers whether or not their child had undergone any additional surgeries since the initial surgery to repair the gastroschisis. Fifteen (25.86%) mothers responded that their children had required at least one other surgery. Seven of these fifteen (46.67%) mothers indicated that two additional surgeries were performed on their children, while one mother reported three additional surgeries for her child.

Development of the Gastroschisis Cases:

Physical

The children's current ages ranged from 3 years to 12 years, with the median age being 5.5 years (IQR = 4-8). Half of our population of gastroschisis cases were born between 2006 and 2008. Dates of birth, current heights, and current weights, as reported on the questionnaires, were used to evaluate growth of the children born with gastroschisis. The median percentile for current heights of the children born with gastroschisis was 59.5th percentile (IQR = 18.5-82.5%), while the median percentile for current weights of the children born with gastroschisis was 64th percentile (IQR = 30.5-87.5%). Of note, however, three of the children in our population were at less than the 3rd percentile for weight only, and five were at less than the 3rd percentile for height only. All eight of these children were between the ages of four and six years old. No children were noted to be at less than the 3rd

percentile for both height and weight. All of the three children that were below the 3rd percentile for weight were males. Regarding the five children who were below the 3rd percentile for height, three were female and two were male.

On the questionnaire, mothers were also asked to compare their children's current sizes to the sizes of other children that are the same ages by answering a multiple choice question. Three (5.17%) mothers indicated that their child was "much smaller than other children that are the same age." Twelve (20.69%) mothers indicated that their child was "only slightly smaller than other children that are the same age." Twenty-five (43.10%) mothers said that their child was "the same size as other children that are the same age." Eleven (18.97%) mothers viewed their child as "only slightly bigger than other children that are the same age." Lastly, four (6.90%) mothers stated that their child was "much bigger than other children that are the same age." The remaining three mothers did not answer this question.

When comparing the children's growth percentiles to parental perceptions of size, we found that only one male who was at less than the 3rd percentile for weight was reported to be "much smaller than other children the same age" by his mother. The mothers of the remaining two males who were at less than the 3rd percentile for weight did not answer the question regarding the size of their child. Three of the children that were at less than the 3rd percentile for height (one male and two females) were reported to be "the same size as other children the same age" by their mothers, while two children (one male and one female) were reported to be "only slightly smaller than other children the same age." Two children that were on their growth curves for both height and weight were describes as "much smaller than other the children the same age" by their mothers.

In addition, the questionnaire asked mothers to indicate how often their child felt tired using a Likert scale that included the following responses: never, sometimes, often, most of the time, and always. Overall, twenty-four (41.38%) mothers chose “never,” thirty-one (53.45%) mothers chose “sometimes,” one (1.72%) mother chose “often,” and two (3.45%) mothers did not answer this question. Because younger children may not be able to express fatigue, counts for children that are at least six years old were evaluated separately as well. For this subset of the study population, which included twenty-nine cases, twelve (41.38%) mothers described their child as never feeling tired, sixteen (55.17%) mothers described their children as sometimes feeling tired, and one (3.45%) mother stated that her child felt tired often.

Educational

Grade levels for the fifty-eight children were obtained from the questionnaire. Eight (14.04%) children were not currently attending school. Ten (17.24%) children were in preschool and 3 (5.26%) children were in pre-kindergarten. Eight (14.04%) children were reported to be in kindergarten. Twenty-one (36.21%) children were in elementary school. Seven (12.07%) children were attending middle school at the time of the study. One (1.72%) mother did not indicate the current grade level of her child.

In order to assess scholastic performance, the study questionnaire asked mothers to indicate the amount of additional help with schoolwork received by their children from teachers or tutors. This was a multiple choice question that included the following answer choices: never, 1-3 hours per week, 4-6 hours per week, 7-10 hours per week, and more than 10 hours per week. For the thirty-seven children who were reported to be in kindergarten,

elementary school, or middle school, the majority (64.86%) never received additional assistance from teachers or tutors. Eight (21.62%) of the children were reportedly receiving 1-3 hours of additional help per week. Three (8.11%) children received 4-6 hours of additional help per week. Two (5.41%) mothers did not provide an answer to this question.

Additionally, the study questionnaire aimed to assess the frequency of learning disability diagnoses in children born with gastroschisis. Forty-seven (81.03%) mothers indicated that their child had not been diagnosed with a learning disability, while ten (17.24%) mothers stated that their child had been diagnosed with a learning disability. Of the ten children diagnosed with a learning disability, two were diagnosed with multiple learning disabilities. One (1.72%) mother did not indicate whether or not her child had been diagnosed with a learning disability. Types of learning disabilities reported in this study population are shown in Table 5.

Table 5: Learning Disability Diagnoses in the Study Population

Learning Disability Diagnoses	Frequency
ADD	6
Autism Spectrum Disorder	1
Developmental Delay	1
Dyslexia	0
Intellectual Disability	2
"Other:"	4
Possible ADD	1
Speech/Language Delay	3

Because the prevalence of learning disabilities differs between full-term and preterm infants in the general population, the full-term and preterm gastroschisis cases were analyzed separately. Of the twenty full-term gastroschisis cases, seventeen (85.0%) were reported to have no learning disability, while three (15.0%) were reportedly diagnosed with a learning disability. A binomial probability test was used to compare the prevalence of diagnosed learning disabilities in our population of full-term gastroschisis cases to the prevalence of approximately 3% in the general population, as reported by the U.S. Census Bureau [46]. The prevalence of learning disabilities in the full-term gastroschisis population was significantly higher than that in the general population ($p = 0.021$).

Similarly, of the thirty-seven preterm gastroschisis cases, seven (18.92%) children were diagnosed with a learning disability, while thirty (81.08%) children had no learning disability diagnosis. Again, a binomial test was used to compare the prevalence of learning disability diagnoses in the preterm gastroschisis population to the prevalence of about 7.7% in preterm infants, as reported in a 2011 study by the Centers for Disease Control Prevention and a 2009 study by van Baar et al. [47, 48]. The preterm gastroschisis population had a significantly higher prevalence of learning disabilities than the preterm infants ($p = 0.021$).

Because learning disabilities are most often diagnosed in children attending school, the subset of gastroschisis cases currently in kindergarten, elementary, or middle school, was further analyzed. When considering only the thirty-six children currently in kindergarten, elementary school, or middle school, twenty-eight (77.78%) did not report a learning disability diagnosis, while eight (22.22%) did report a learning disability diagnosis. Three of the eight school-attending children (8.3% of all school-going children) with diagnosed learning disabilities were born at term, while the remaining five (13.9% of all school-going

children) were born preterm. Binomial tests were used to determine whether or not the prevalences of learning disability diagnoses were significantly different between full-term gastroschisis cases that were currently in school and children in the general population, as well as between preterm gastroschisis cases that were currently in school and other previously preterm infants in the general population. Prevalences of learning disability diagnoses were significantly higher in both school-going populations, with p-values of 0.011 for the full-term gastroschisis cases and 0.016 for the preterm gastroschisis cases.

Food Intolerances:

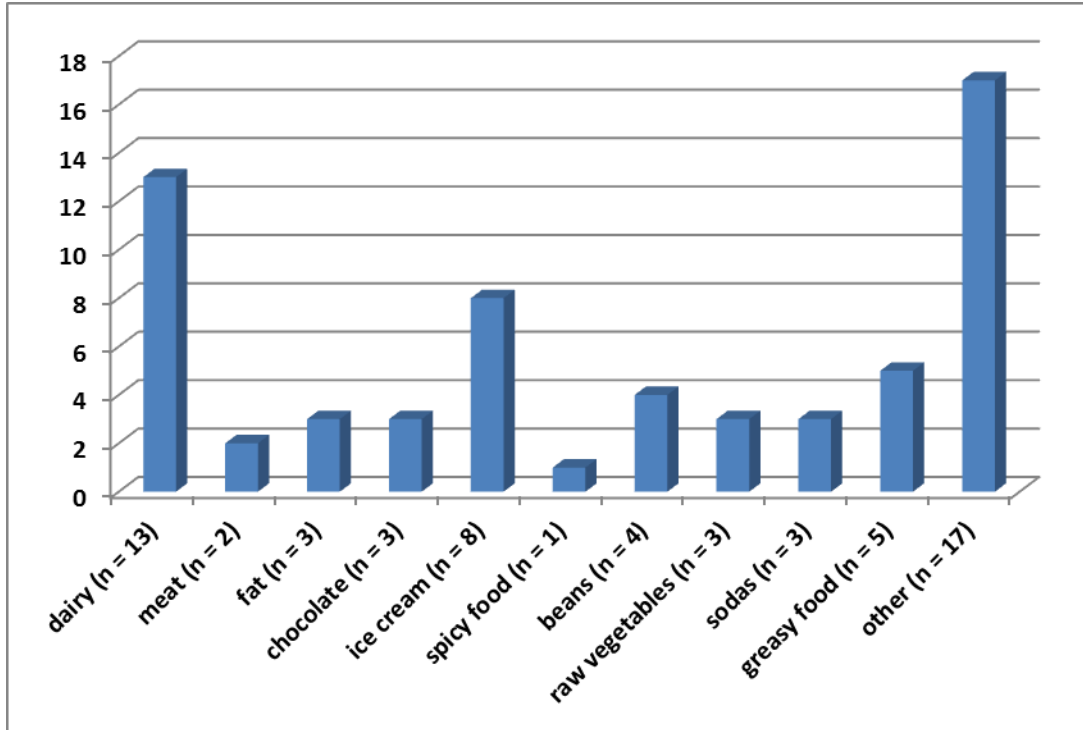
Questions were included on the study questionnaire to determine whether or not children born with gastroschisis experience frequent discomfort after eating certain foods. Parents were asked to answer the Likert scale question, "my child is unable to eat certain foods without feeling uncomfortable:" with the following answer choices: never, sometimes, often, most of the time, and always. Because young children may not be able to express discomfort after eating certain foods, responses to this question were described only for children 6 years and older. Of the fifty-eight children in the study population, twenty-nine were at least 6 years old. Responses concerning these twenty-nine children are presented in Table 6.

Table 6: Frequency of Food-Related Discomfort in Children At Least 6 Years Old

Frequency of Discomfort Due to Certain Food in Children 6 years and Older	
Reported Frequency of Discomfort	n (%)
Never	19 (65.52%)
Sometimes	7 (24.14%)
Often	1 (3.45%)
Always	2 (6.90%)

Question fifteen on the questionnaire asked parents to circle or list any foods that cause their children to feel uncomfortable. The following categories were given as options: milk/dairy, meat/protein, fat/fatty foods, chocolate, ice cream, chips, spicy foods, gluten, beans, raw vegetables, sweetener (e.g. sorbitol), sodas, greasy/fried foods, and other. Parents were given space to write in any "other" answers. Of the fifty-eight children included in this study, thirty-nine (67.24%) were reported to have no food intolerances. Twelve (20.69%) children reportedly had 1-2 food intolerances, while seven (12.07%) children had more than two food intolerances. Food categories that were indicated as causing discomfort in this study population are represented in Figure 4. Because dairy and ice cream products were frequently reported as causing discomfort, a binomial test was used to compare the prevalences of lactose intolerance in various ethnicities in the general population, based on a 2010 report released by the Agency for Healthcare Research and Quality [49], to the prevalence of dairy and ice cream intolerance in this study population. Neither the non-Hispanic children nor the Hispanic children in the study population had significantly increased prevalences of lactose intolerance ($p = 0.999$ and $p = 0.955$, respectively).

Figure 4: Foods Causing Discomfort In Gastroschisis Cases



Gastrointestinal Issues:

In addition, the study questionnaire contained questions aimed at assessing the frequency of select gastrointestinal issues in children born with gastroschisis. Parents were asked to indicate how often their children complained of abdominal pain, had problems with reflux, had problems with diarrhea, and had problems with constipation. The Likert scale for these issues contained the following options: rarely, a few times a year, a few times a month, a few times a week, and everyday. The frequencies of these issues in this study population are presented in Table 7.

Table 7: Reported Frequencies of Abdominal Pain, Reflux, Diarrhea, and Constipation in Gastroschisis Cases

	Abdominal Pain (n)	Reflux (n)	Diarrhea (n)	Constipation (n)
Rarely	32	47	35	30
Few Times Yearly	10	3	11	12
Few Times Monthly	9	2	7	4
Few Times Weekly	2	1	2	6
Everyday	3	1	0	3
Missing Responses	2	4	3	3

Two other similar Likert scale statements included in the questionnaire were: "my child feels anxious when there is not a restroom nearby:" and "my child complains of not being hungry:” The Likert scale for these statements consisted of the following options: never, sometimes, often, most of the time, and always. These questions aimed to assess frequency of complaints or anxieties that may be secondary to gastrointestinal issues. The responses to these statements, when considering children six years and older only, are presented in Table 8. Children under the age of six were excluded from the analysis because young children are unlikely to express these feelings of anxiety or lack of hunger.

Table 8: Frequency of Lack of Hunger and Feeling Anxious Without a Nearby Restroom in Children at Least 6 Years Old

Frequency of Feeling. . .	Not Hungry *	Restroom Anxiety
	n (%)	n (%)
Never	19 (70.37%)	22 (75.86%)
Sometimes	7 (25.93%)	6 (20.69%)
Most of the Time	1 (3.7%)	0 (0.0%)
Always	0 (0.0%)	1 (3.45%)

* 2 respondents did not answer

The questionnaire also included a multiple choice question that asked parents how often their children missed school due to gastrointestinal problems or discomfort. The answer choices were as follows: 1-2 times per week, 1-2 times per month, once every 2-3 months, once every 6 months, and never. Responses to this question were included in the descriptive analysis only if the child was currently attending school. Most mothers (86.11%) of the 36 school-going children reported that their children never missed school because of gastrointestinal issues. The responses to this question are presented in Table 9.

Table 9: Frequency of Missing School Due to Gastrointestinal Issues in School-age Children

Frequency of Missing School	n (%)
1-2 Times/Week	0 (0.0%)
1-2 Times/Month	2 (5.56%)
Once Every 2-3 Months	0 (0.0%)
Once Every 6 Months	3 (8.33%)
Never	31 (86.11%)

Amount of Care Required from Parents:

One multiple choice question on the study questionnaire asked parents how much time and care their child needed in comparison to other children the same age. The majority of parents (70.69%) reported that their child required the same amount of time and care as other children that are the same age. The responses to this question are displayed below in Table 10.

Table 10: Amount of Time and Care Needed by Children Born with Gastroschisis

Time and Care Required from Parents	Frequency of Responses n (%)
Much More	2 (3.45%)
Slightly More	9 (15.52%)
Same Amount	41 (70.69%)
Slightly Less	2 (3.45%)
Much Less	1 (1.72%)
Missing Responses	3 (5.17%)

Self-esteem Related to the Surgical Scar:

Because self-esteem is often affected by physical appearance, and children with repaired gastroschisis have noticeable scars, the questionnaire contained a Likert scale question that aimed to assess how often children with repaired gastroschisis felt self-conscious about their surgical scars. The Likert scale for this question contained the following options: never, sometimes, often, most of the time, and always. Responses to this

question are presented in Table 11; however, only responses regarding children six years or older were evaluated to ensure that the sample included children that were old enough to have potentially developed feelings of self-consciousness related to their scars.

Table 11: Frequency of Feeling Self-Conscious About the Scar in Children 6 Years and Older

Frequency of Feeling Self-Conscious	n (%)
Never	15 (51.72%)
Sometimes	10 (34.48%)
Often	0 (0.0%)
Most Times	3 (10.34%)
Always	1 (3.45%)

Reported Use and Frequency of Medications:

The study questionnaire also aimed to identify medications commonly used by patients born with gastroschisis. A list of medications was provided, and parents were asked to mark whether their children required each medication daily, weekly, as needed, or never. The medications that were listed included: vitamins, allergy medication, reflux medication, diarrhea medication, nausea/vomiting medication, medication for stomach aches, medication for stomach ulcers, medication for constipation, medication for gas/bloating, medication for lactose intolerance, medication for ulcerative colitis/Crohn's disease/Irritable Bowel Syndrome, medication for ADD/ADHD, asthma medication, and general pain relievers. The reported frequencies of using each of these medications are presented in Table 12 on the following page.

Table 12: Reported Medication Use in the Gastroschisis Population

	Never (n)	As Needed (n)	Weekly (n)	Daily (n)	Missing Responses (n)
Vitamins	22	12	1	21	2
Allergy Medicine	26	23	2	4	3
Reflux Medicine	51	4	0	0	3
Diarrhea Medicine	41	14	0	0	3
Nausea Medicine	47	9	0	0	2
Stomach Ache Medicine	42	11	2	0	3
Stomach Ulcer Medicine	53	1	0	1	3
Constipation Medicine	40	14	1	1	2
Gas/Bloating Medicine	49	4	1	1	3
Lactose Intolerance Medicine	52	3	0	0	3
Ulcerative Colitis Medicine	54	1	0	0	3
ADD Medicine	51	1	0	2	4
Asthma Medicine	47	6	0	2	3
Pain Relievers	10	44	1	0	3

Specialists Frequently Seen:

A Likert scale was also included on the study questionnaire to determine the frequency of seeing various specialists within the gastroschisis population. Parents were asked to indicate whether their children had seen a given specialist once, twice, three times, more than three times, or never. The specialists that were listed were as follows: audiologists, cardiologists, developmental pediatricians, otolaryngologists, gastroenterologists, geneticists, nephrologists, neurologists, pulmonologists, pediatric surgeons, social workers, physical therapists, and occupational therapists. An "other" option was also given for parents to list another specialist. The results of these responses are presented in Table 13.

Table 13: Specialists Seen and Frequency of Visits

	Never (n)	Once (n)	Twice (n)	Three Times (n)	> Three Times (n)	Missing Responses (n)
Audiologist	27	13	7	1	5	5
Cardiologist	44	5	3	0	1	5
Developmental Pediatrician	44	2	2	2	4	4
Otolaryngologist	29	12	6	1	5	5
Gastroenterologist	21	11	9	6	7	4
Geneticist	51	22	1	0	0	4
Nephrologist	53	0	0	1	1	3
Neurologist	49	3	1	1	1	3
Pulmonologist	46	2	1	1	1	7
Pediatric Surgeon	20	16	7	3	10	2
Social Worker	49	1	1	2	1	4
Physical Therapist	47	1	0	2	5	3
Occupational Therapist	47	2	0	2	2	5
Other	2	0	1	0	2	0

Audiology visits were common in this population of gastroschisis cases. Fifty-three mothers indicated the number of times that their children had seen an audiologist. These responses were dichotomized into two groups: those that indicated seeing an audiologist less than three times and those that indicated seeing an audiologist three or more times. The threshold for the dichotomization was selected based on the assumption that those that indicated seeing an audiologist at least three times were assumed to have some type of hearing impairment that was the underlying reason for the frequent audiology visits. Binomial tests were used to determine whether, according to ethnicity, the prevalence of presumed hearing impairment in this study population was significantly higher than the prevalence of hearing impairment in children ages 6-19 in the United States as indicated by the National Health and Nutrition Examination Survey II [50]. Results showed that non-Hispanic white children older than six years old and born with gastroschisis have a presumably higher prevalence of hearing impairment than 6-19 year old non-Hispanic white children in the general population ($p < 0.0005$), (Table 14). Hispanic children over the age of six and born with gastroschisis were not found to have a significantly higher prevalence of presumed hearing impairment than the general Hispanic population. However, in this study population, there were only nine Hispanic children over the age of six whose mothers indicated number of visits to an audiologist, and none of these reported at least 3 visits to an audiologist. In the African American children in our population, we did not observe a significantly increased prevalence of presumed hearing impairment; however, similar to the Hispanic population, the sample size in this subset was limited to only two African American children over the age of six whose mothers indicated how often their children had

seen an audiologist. Neither of these mothers reported 3 or more audiology visits for their child.

Table 14: Binomial Test Results Comparing Prevalences of Hearing Impairment* in Gastroschisis Cases to Prevalence of Hearing Impairment in the General Population, By Ethnicity

Ethnicity (Children ≥ 6 Years Old Only)	Presumed Hearing Impairment* in the Gastroschisis Cases	Prevalence of Hearing Impairment in the General Population	p-value
Non-Hispanic White	28.57% (4/14)	0.38%	p < 0.0005
African American	0.0 % (0/2)	0.78%	p = 1.00
Hispanic	0.0% (0/9)	0.60%	p = 1.00

*Hearing impairment was presumed to be present in individuals that saw an audiologist at 3 or more times

Of note, visits to audiologists and otolaryngologists (ENT) were not fully correlated among the children in this study. Of the thirty-one children in this study who reported seeing an ENT or audiologist at least one time, eleven children saw an audiologist more often than an ENT, and eight children who saw an audiologist never saw an ENT. Additionally, nine children saw an ENT more often than an audiologist, and six children who saw an ENT never saw an audiologist.

Because learning disabilities were identified as a significant morbidity in children born with gastroschisis, we used the Fisher's exact test to determine whether the increased

frequency of audiology visits in our study population was significantly associated with diagnosis of a learning disability, as children suspected of having a learning disability are often screened by audiologists. Based on the results of Fisher's exact test, we found that the frequency of audiology evaluations was not significantly associated with diagnosis of a learning disability ($p = 1.00$).

Free Response Questions:

Four free response questions were included on the questionnaire. Statistical analyses were not performed on the responses to these questions, although trends were noted. The first question asked parents to describe what they consider to be "good quality of life." Forty-six mothers provided responses to this question. Of these forty-six, the majority (54%) indicated that a "good quality of life" was contingent on good health. Also, of note, approximately thirty-three percent of respondents mentioned that happiness was a contributing factor to overall quality of life. Nine mothers (19.6%) also considered a stable home environment and supportive family to be important to having a "good quality of life."

The second open-ended question asked parents to describe their child's quality of life. Fifty-one mothers answered this question. Approximately half of these mothers (53%) described their child's quality of life using words such as "good," "great," "excellent," "wonderful," or "fantastic." Over one-fifth of the mothers stated that they considered their child to be normal or on par with other children the same age. Eight mothers (16%) noted that their children experienced health complications, physical limitations, or self-esteem issues secondary to the gastroschisis.

The third free response question asked mothers what advice they would offer to parents who were first receiving a diagnosis of gastroschisis for their child. Fifty-four mothers responded to this question. Overall, the majority (72%) encouraged other parents to maintain an optimistic outlook and expect a positive outcome. These mothers stated that, although it is difficult for parents to receive a diagnosis of gastroschisis initially, it becomes easier over time. Fourteen (25.9%) of the women who answered this question also encouraged parents to take comfort in the knowledge of the medical community and to trust the doctors who would be taking care of the child, saying that these doctors are very experienced and equipped to manage the child. Nine mothers emphasized the importance of having faith and trusting God to take care of the child. Interestingly, although 80% of the mothers who broached the issue of researching the diagnosis (n=10) were for it, the remainder (n=2) cautioned parents against doing additional research.

Lastly, the questionnaire included an open-ended response section where mothers could write comments for the researchers involved in this study. Twenty-nine mothers provided comments. Approximately forty-five percent of comments included in this section consisted of statements or questions regarding the cause, prognosis, or recommended management and treatment of issues their children had experienced. Also, seven (24.1%) mothers expressed gratitude to the researchers for looking into gastroschisis in more depth.

PedsQL 4.0 Scales Scores:

The PedsQL 4.0 scales were completed and returned by fifty-five mothers. The PedsQL 4.0 scales were used to determine the physical quality of life score, psychosocial quality of life score, and total quality of life score for each child. The median physical score

for this study population was 96.88 (IQR=90.63-100). The median psychosocial score was 86.67 (IQR=75.0-95.0). The median total quality of life score for this population was 91.3 (IQR=78.26-95.65).

The mean total score of 87.02 (SD 12.78) in this study population was significantly higher than the previously reported mean score of 82.70 (SD 15.40) in reportedly "healthy" children ($p = 0.019$) [51]. The mean physical quality of life score of 93.55 (SD 10.51) in our population was also significantly higher than the mean physical score of 84.48 (SD 19.51) in the population of children reported to be "healthy" ($p = 0.0003$). Lastly, although the mean psychosocial score of 83.20 (SD 16.22) in the gastroschisis cases was slightly higher than the mean psychosocial score of 81.65 (SD 15.22) in "healthy" children, this difference did not achieve statistical significance ($p = 0.228$) [51]. Overall, the children with gastroschisis did not have qualities of life that are worse than healthy children.

Similarly, PedsQL 4.0 scores in our population were compared to PedsQL 4.0 scores in children with other chronic conditions. Our population of children born with gastroschisis had significantly higher PedsQL 4.0 total, physical, and psychosocial scores than children with other chronic conditions such as asthma ($p < 0.0005$), gastrointestinal conditions ($p < 0.0005$), and cancer ($p < 0.0005$) [51].

Factors Associated with PedsQL 4.0 Scores:

Wilcoxon rank sum tests and Kruskal-Wallis tests were used to determine whether or not any of the factors identified through the questionnaire or data obtained from the TBDR were significantly associated with the physical, psychosocial, or total scores of the PedsQL 4.0. In statistical analysis, the reported frequencies of variables, such as experiencing

specific health issues or seeing specialists, were dichotomized into "never" and "ever" because the sample sizes in the individual categories of the Likert scale were very small.

Physical PedsQL 4.0 Scores

Variables were analyzed to determine factors that significantly affect physical quality of life. Factors that were significantly associated with PedsQL 4.0 physical scores were feeling tired, feeling uncomfortable after eating certain foods, having intolerances to certain food groups, having abdominal pain, having reflux issues, constipation, taking medications for reflux, taking medications for stomach aches, taking medications for constipation, seeing an otolaryngologist, seeing a social worker, and having additional surgeries (Table 15).

Table 15: Variables Significantly Associated with Physical PedsQL 4.0 Scores

Variable	p-value
Feeling Tired	0.003
Experiencing Food Discomfort (Age \geq 6)	0.045
Having Food Intolerances (None, 1-2, or >2)	0.016
Abdominal Pain	0.0006
Reflux	0.0003
Constipation	0.012
Taking Reflux Medication	0.015
Taking Stomach Ache Medication	0.006
Taking Constipation Medication	0.005
Seeing Otolaryngology	0.008
Seeing Social Work	0.003
Having Additional Surgeries	0.041

Psychosocial PedsQL 4.0 Scores

Factors significantly associated with the psychosocial PedsQL 4.0 scores were feeling tired, not feeling hungry, taking medications for stomach aches, seeing a developmental pediatrician, having seen a social worker, seeing a physical therapist, having a learning disability, receiving tutoring, missing school due to gastrointestinal issues, and requiring more time and care from parents than other children the same age. The p-values for these significant factors are displayed in Table 16.

Table 16: Variables Significantly Associated with Psychosocial PedsQL 4.0 Scores

Variable	p-value
Feeling Tired (Age \geq 6)	0.015
Feeling Not Hungry (Age \geq 6)	0.003
Taking Stomach Ache Medication	0.009
Seeing a Developmental Pediatrician	0.027
Seeing Social Work	0.016
Seeing a Physical Therapist	0.027
Having a Learning Disability	0.001
Receiving Additional Tutoring	0.024
Missing School Due to GI Issues	0.035
Requiring Additional Care	0.040

Total PedsQL 4.0 Scores

Variables that were found to significantly affect the total PedsQL 4.0 scores of children born with gastroschisis were feeling tired, not feeling hungry, taking medications for stomach aches, seeing a developmental pediatrician, having seen a social worker, having a learning disability, requiring extra tutoring, missing school due to gastrointestinal issues, and requiring more time and care from parents than other children the same age. Table 17 displays the p-values for these factors.

Table 17: Variables Significantly Associated with Total PedsQL 4.0 Scores

Variable	p-value
Feeling Tired (Age ≥ 6)	0.006
Feeling Not Hungry (Age ≥ 6)	0.002
Taking Stomach Ache Medication	0.004
Seeing a Developmental Pediatrician	0.022
Seeing Social Work	0.026
Having a Learning Disability	0.001
Receiving Additional Tutoring	0.049
Missing School Due to GI Issues	0.020
Requiring Additional Care	0.032

Factors Associated with Morbidities in Gastroschisis Patients

Wilcoxon rank sum tests and Kruskal-Wallis tests were used to determine whether or not morbidities in gastroschisis patients are affected by any demographic factors (Table 13). Gender of the gastroschisis case was found to be significantly associated with certain physical and psychological issues in this study population. Feeling self-conscious about the surgical repair scar was associated with being female. Anxiety due to not having a restroom nearby was also associated with being female. In addition, taking medications for gas or bloating was found to have an association with being a female. Being male was significantly associated with an increased frequency of seeing an audiologist when considering all fifty-three children for whom frequency of audiology visits was reported. However, after stratifying frequency of audiology visits by ethnicity (since ethnicity on its own was associated with prevalence of hearing impairment), gender of the gastroschisis case was not significant. P-values for the above associations are shown in below in Table 18.

Tables 18: Statistically Significant Factors Associated with Morbidities in Gastroschisis Cases

Variable	p-value
Being Female and Feeling Self-Conscious About the Scar	0.022
Being Female and Restroom Anxiety	0.026
Being Female and Taking Medications for Gas/Bloating	0.021
Being Male and Seeing Audiologists	0.017
White Males Seeing Audiology	0.126
African American Males Seeing Audiology	0.157
Hispanic Males Seeing Audiology	0.103

Discussion:

This study aimed to describe the long-term quality of life and morbidities in children who were born with gastroschisis. Additionally, we attempted to identify factors that were significantly associated with decreased PedsQL 4.0 quality of life scores and with morbidities observed in this population.

Quality of Life:

When considering quality of life as measured by the PedsQL 4.0 scales, it appears that this study population of children born with gastroschisis has a mean total quality of life score (87.02) that is significantly higher than the mean score of 82.70 in healthy children ($p = 0.019$) [51]. The mean physical quality of life score of 93.55 in our population is also significantly higher than the mean physical score of 84.48 in healthy children ($p = 0.0003$). However, the mean psychosocial score of 83.20 in the gastroschisis cases, although greater than the mean psychosocial score of 81.65 in healthy children, is not significantly higher ($p = 0.228$) [51]. Based on these reported scores, it appears that quality of life in children with gastroschisis is not worse than the quality of life in healthy children. Similarly, in comparison to children with chronic conditions such as asthma, gastrointestinal conditions, and cancer, our population of children born with gastroschisis had significantly higher PedsQL 4.0 physical ($p < 0.0005$), psychosocial ($p < 0.0005$), and total ($p < 0.0005$) scores [51]. Therefore, it seems that in contrast to children who suffer from other childhood conditions, the quality of life during the first 12 years in children with repaired gastroschisis is significantly better. Furthermore, when total PedsQL 4.0 scores are decreased in gastroschisis cases, the decreased total scores appear to be due to lower psychosocial scores rather than to lower physical scores. This observation suggests that, once repaired, children

with gastroschisis experience few major physical limitations; however, complications secondary to the gastroschisis or its surgical repair may negatively influence an individual's psychosocial health more than they influence physical health.

Based on our study population, it appears that overall quality of life was good for most gastroschisis patients. However, when patients experienced issues such as feeling tired ($p = 0.006$), not feeling hungry ($p = 0.002$), taking stomach ache medications ($p = 0.004$), having a learning disability ($p = 0.001$), requiring additional tutoring ($p = 0.049$), missing school due to gastrointestinal issues ($p = 0.020$), or requiring additional care from parents ($p = 0.032$), these patients' total quality of life scores were impacted. Similarly, lower total quality of life scores were observed among children who reported seeing developmental pediatricians ($p=0.022$) or social workers ($p=0.026$).

Likewise, factors associated with psychosocial quality of life were identified. In our study, we found decreased psychosocial quality of life to be associated with feeling tired ($p = 0.015$), not feeling hungry ($p = 0.003$), taking stomach ache medications ($p = 0.009$), missing school due to gastrointestinal complications ($p = 0.035$), requiring additional care from parents ($p = 0.040$), having a learning disability ($p = 0.001$), and receiving additional tutoring ($p = 0.024$). Again, since children seeing a developmental pediatrician ($p = 0.027$), social worker ($p = 0.016$), or a physical therapist ($p = 0.027$) probably have some health issue that is being addressed by these professionals, it was not surprising that these factors were found to be associated with lower psychosocial quality of life scores. Of note, nine of the ten factors that were found to be associated with psychosocial quality of life were also found to have a significant association with total quality of life, illustrating the effect of psychosocial quality of life on total quality of life in our population. Further study into the

underlying etiologies for these factors with the goal of identifying possible means to limit their effect on quality of life is warranted.

Lastly, factors that were significantly associated with physical PedsQL 4.0 scores were identified. Significant associations between physical PedsQL 4.0 scores and feeling tired ($p = 0.003$), experiencing discomfort related to specific foods ($p = 0.045$), complaining of abdominal pain ($p = 0.002$), taking medication for stomach aches ($p = 0.006$), experiencing reflux ($p = 0.011$), taking medication for reflux ($p = 0.015$), experiencing constipation ($p = 0.010$), taking medication for constipation ($p = 0.005$), having intolerance to certain foods ($p = 0.016$), and undergoing additional surgeries ($p = 0.041$) were found to be associated with lower quality of life scores. Also, seeing an otolaryngologist ($p = 0.046$) or a social worker ($p = 0.047$) was marginally associated with physical PedsQL 4.0 scores. Many of these factors are related to gastrointestinal pain or discomfort, which may limit an individual's physical functioning and thus physical quality of life. Further research is needed to clarify the associations between physical quality of life and the issues described above.

Morbidities:

In respect to the morbidities observed in our population, learning disabilities were found to occur at a significantly higher rate in children born with gastroschisis than in children in the general population. Full-term ($p=0.021$) and preterm ($p=0.021$) gastroschisis cases were found to have learning disabilities significantly more often than full-term and preterm infants in the general population. This finding remained significant when considering only the children who were currently attending kindergarten, elementary school,

or middle school. The association between gastroschisis and an increased risk of learning disabilities has not been reported in previous studies. A 2012 study by Gorra et al. evaluated the 2- year neurodevelopmental outcomes in children born with gastroschisis and did not find a statistically significant rate of delayed development in the gastroschisis cases compared to gestational age-matched and birth weight-matched NICU controls with no surgical history [52]. Gorra et al. concluded that, although the gastroschisis cases appeared to have an increased rate of developmental delay compared to the general population, the delay observed in the gastroschisis cases was a result of premature delivery and unrelated to the actual birth defect. However, Gorra et al. did not follow patients through school age when the majority of learning disabilities are diagnosed. In contrast, our study found that children born with gastroschisis have a statistically significant increased risk to develop learning disabilities in comparison to the general population, regardless of gestational age at birth. There was no statistically significant association between learning disability diagnoses and the type of surgical repair of gastroschisis ($p = 0.706$) or whether the child underwent additional surgeries ($p = 0.596$). Future studies should be done to identify potential causes of the increased risk for learning disabilities in children born with gastroschisis as anticipatory guidance for parents and physicians regarding the increased prevalence of learning disabilities may lead to earlier diagnosis and intervention and improved outcome if this finding is replicated.

In addition, based on the reported frequencies of visits to an audiologist in our study population, it appears that non-Hispanic white children born with gastroschisis may be at an increased risk of hearing impairment (defined as three or more visits to an audiologist in our dataset). Lee et al. reported that, based on 7,888 children between the ages 6 to 19 years old

who were included in the National Health and Nutrition Examination Survey (NHANES) II and Hispanic Health and Nutrition Examination Survey (HANES) studies, the prevalence of hearing impairment (defined as 31+ dBHL in the better ear) was 0.38% in the non-Hispanic white population. This prevalence rate was significantly lower than the rate of 28.6% (4 out of 14 children) observed in our population of non-Hispanic white children ages 6 and older ($p < 0.0005$). This finding in our study sample was independent of the presence of learning disabilities, with 25% of children with learning disabilities and 30% of children without learning disabilities reporting at least three visits to an audiologist ($p = 0.857$). However, it should be noted that we hypothesized that three or more visits to an audiologist could be taken as a proxy for hearing loss. The questionnaire did not inquire specifically about clinically diagnosed hearing loss or the reason for visiting an audiologist. Further research is required to elucidate the relationship between gastroschisis and hearing loss.

Also, regarding morbidities, growth restriction did not appear to be a significant issue in the majority of our gastroschisis cases, as the median percentiles for current weights and heights were 64th and 59.5th percentiles, respectively. These percentiles are based on the reported weights of forty-eight children and the reported heights of forty children. Of note, however, eight of the children in our population were at less than the 3rd percentile for weight or height only. All eight of these children were between the ages of four and six years old. Based on the findings in our population, it appears that the few children who experienced growth delay were relatively young. Previous studies have also reported growth to be an issue in up to 33% of gastroschisis patients [24, 33, 34]. In a study by South et al., approximately one-third of children born with gastroschisis were below the 10th percentile at 16 to 24 months [24]. The authors of this study state that the large number of

patients with growth restriction in their study may be due to the young age at follow-up, considering that many of the children evaluated were born premature or were small for gestational age at birth and may not have caught up by 24 months of age. This assumption by South et al. seems to be borne out in our dataset, where 24% of the children aged 3 years or older were below the 10th percentile for height or weight. Additionally, South et al. hypothesize that their large number of children with growth delay may be reflective of recent advances in medical care that allow for increased survival of severe gastroschisis cases already at an increased risk for growth delay [24].

Need for Additional Counseling:

While the data suggests that children with gastroschisis may be at risk for hearing impairment and learning disabilities, their total quality of life scores are not decreased from healthy children in the population, and in fact were significantly increased from expected. The answers provided to the free response questions on the study questionnaire reaffirm the observation that being born with gastroschisis does not drastically affect a child's quality of life for the majority of children. Most of the mothers who answered the question "How would you describe the quality of life of your child with gastroschisis?" said that they considered their child's quality of life to be good or the same as other children the same age. For example, one mother said that her daughter is "[. . .] very good in school, holds an A average. She is happy in her own skin and loves others. She has a good quality of life." Similar answers were given by over half (53%) of other mothers.

Interestingly, however, when leaving comments for the researchers of this study, many of the mothers had questions regarding the long-term prognosis and causes of

gastroschisis, as well as questions about additional issues experienced by their child. One mother wrote the following comment:

“Interested to know about damage to abdominal wall muscles. What happens to them? My son's seem to be misshapen around his scar. Doesn't seem to cause discomfort or lack of core strength, but I want to know are they different because of the initial surgery? I would love a follow-up if possible, as well as to participate in any other way to help with families of gastroschisis babies. Please see contact info if needed: [. . .]”

Because of the uncertainty reflected in the responses of many mothers (44.8%), it appears that the counseling given to parents receiving a diagnosis of gastroschisis for their child does not address the long-term questions that arise. Although mothers who completed our questionnaire were mostly optimistic in the advice they would offer to the parents of children newly diagnosed with gastroschisis, it seems as though they are uncertain about the appropriate management of and long-term prognosis for gastroschisis. Therefore, health care professionals working with families with gastroschisis may wish to incorporate discussions about long term follow-up into their clinical care plan.

Finally, there appears to be a small, but relevant, subset of children with gastroschisis who experience more significant complications, such as frequent abdominal pain, constipation, incontinence, and low self-esteem. Mothers of 16% of our sample indicated that their children experienced health complications or self-esteem issues that impacted their quality of life. One mother indicated that her daughter had a good overall quality of life, but that she often felt embarrassed about her surgical scars. This mother stated that she was considering plastic surgery as a future option for her daughter. Similarly, one mother reported that her son had a “fair” quality of life, but that he had “struggled with learning disabilities, vision, low self-esteem, and a few others.” Previous studies examining long-term survivors of gastroschisis have also found these patients to be affected by low

self-esteem and gastrointestinal complications [30, 31]. Although limited in sample size, the Koivusalo et al. study also found 12% of its population to be affected by self-esteem issues [31]. Recurrent gastrointestinal complications were observed in 7.1% of the population in the Henrich et al. study [30]. It might be helpful for families to be advised of the possibility that these factors may arise so that they may seek out appropriate management in an expedient fashion.

Limitations:

The sample size of this study is both a strength and a limitation. Although relatively small (n=58), our sample of gastroschisis cases is still larger than that of any other published study and appears to be representative of the overall population of gastroschisis cases in the TBDR. However, if our response rate had been higher, the power of the study to tease apart subtle differences would have been higher. Therefore, the significant findings in our population warrant further research and validation in larger patient populations before these findings can be labeled as representative of all gastroschisis patients.

Another limitation of our study is that since the TBDR data is only complete through 2008, this study represents only children who survived at least the first three years of life. Additionally, based on our IRB protocol we were not able to obtain information on mortality and therefore are unaware of how many of the 1,112 questionnaires sent out were not returned due to the fact that the child is deceased. Therefore, our population may be biased towards the less severe cases of gastroschisis, and the overall “normal” quality of life observed in our study may not be reflective of quality of life in severe cases.

As with all retrospective questionnaires, parental recall of information from their child's past medical history introduces a limitation to the study. Several of the questions on this study instrument required parents to recall details of delivery or care received by their child shortly after birth. Parents may have been unable to accurately recall aspects of their child's birth or past surgical history, particularly for children who were born several years ago. Additionally, the questionnaire asked parents to report the number of times their children had seen various healthcare specialists. Specialists who were seen only once, or those who were seen several years ago, may have been underreported or forgotten entirely.

Future Directions:

Future studies are needed to reproduce the results of this study in other populations of gastroschisis patients. This study identified a significantly increased risk for learning disabilities and, possibly, hearing impairment in children born with gastroschisis in Texas. Additional studies with larger populations are necessary to verify that these increased risks are representative. Future studies should also aim to identify underlying reasons for the increased risks of learning disabilities and hearing impairment in the gastroschisis population as well as to elucidate the factors that influence the subset of patients with decreased quality of life scores.

Conclusions:

Based on the findings of this study, we conclude that overall, QoL during the first 12-years for children born with gastroschisis is comparable to that of healthy children in the general population and better than that of children with other chronic health conditions.

However, a small subset of the gastroschisis population does suffer from long-term health issues, especially gastrointestinal symptoms. Factors such as feeling tired often, having frequent stomach aches, being unable to tolerate certain foods, dealing with constipation, having a learning disability, and requiring more time and care than peers all appear to be significantly associated with a decreased quality of life for a subset of gastroschisis patients. Most significantly, children with gastroschisis have an increased risk for learning disabilities which may or may not be related to an increased risk for hearing impairment. Health care providers involved in the care of patients with gastroschisis should be aware of these associations and should provide anticipatory guidance to parents receiving counseling for a diagnosis of gastroschisis in their child.

Appendix A: Letter of Invitation:

An Invitation to Participate in a Survey about Health and Quality of Life of Children with Gastroschisis

Dear Parent or Guardian:

You are invited to help with research about the long-term effects of gastroschisis on your child's quality of life and health. The purpose of this research is to better understand the experiences of children born with gastroschisis, so that health professionals can better explain to families like yours what to expect.

This survey is anonymous and optional. The results will be used for research purposes only. It should take less than 20 minutes to complete this survey. Both English and Spanish versions are enclosed and you may use whichever language you prefer.

IF YOU AGREE TO PARTICIPATE, PLEASE FOLLOW THESE DIRECTIONS:

- The person who takes care of the child with gastroschisis should complete all parts of the survey.
- Please answer as completely and honestly as you can.
- You can choose to skip any questions that you do not want to answer, and you can stop taking the survey at any time.
- When you are done, please put the completed forms in the return envelope provided and send it back by mail as soon as possible.

IMPORTANT INFORMATION ABOUT YOUR PRIVACY:

- All answers are confidential.
- Data will be summarized and presented as part of a thesis project at The University of Texas Graduate School of Biomedical Sciences at Houston.
- Although the results of the study may be submitted for publication, your name will not be connected to your responses and your name will not appear in any reports or publications.
- You will not be paid for taking this survey.
- If you choose not to participate, or change your mind after you started participation, you will not lose any of your benefits.
- The Texas Birth Defects Registry is part of the Texas Department of State Health Services that collects information about babies with birth defects to track rates of birth defects, look for causes of birth defects, improve public health, and educate families about available services.

The results of this study may help others learn more about the long-term health complications and quality of life of people with gastroschisis. However, there may be no direct benefit to you for participating in this study. This survey has been approved by the Institutional Review Boards of both the Texas Department of State Health Services and the University of Texas Health Science Center at Houston.

If you have any questions or concerns, please contact Roya Mostafavi, BS at Roya.M.Mostafavi@uth.tmc.edu or by calling 713-500-5760. In addition, if you have questions about your rights as a research subject, you may call the Department of State Health Services Institutional Review Board at 512/776-2202 or toll-free at 1-888-777-5037.

Thank you very much for helping with this important research.

Sincerely,

Roya Mostafavi, BS
UT Health
Genetic Counseling Intern
Principal Investigator

Claire Singletary, MS CGC
UT Health
Director, Genetic Counseling Program
Committee Chair

Karen Moffitt, MPH
Texas Birth Defects
Epidemiology &
Surveillance
Texas Dept. of State
Health Services

Appendix B: Letter of Informed Consent:

INFORMED CONSENT FORM TO TAKE PART IN RESEARCH Long-term Outcome Analysis of Morbidity and Quality of Life in Patients Born with Gastroschisis

HSC-MS-11-0387
TDSHS IRB# 11-065

INVITATION TO TAKE PART

You are invited to take part in a research project called, *Long-term Outcome Analysis of Morbidity and Quality of Life in Patients Born with Gastroschisis*, conducted by Roya Mostafavi, BS, of the University of Texas Health Science Center. For this research project, they will be called the Principal Investigator or PI.

Your decision to take part is voluntary. You may refuse to take part or choose to stop taking part at any time. A decision not to take part or to stop being a part of the research project will not change the services available to you.

You may refuse to answer any questions asked on any forms. This research project has been reviewed by the Committee for the Protection of Human Subjects (CPHS) of the University of Texas Health Science Center at Houston as HSC-MS-11-0387. The study has also been reviewed by the Texas Department of State Health Services as IRB# 11-065.

PURPOSE

The purpose of this research study is to better understand the experiences of children born with gastroschisis, so that health professionals can better explain to families like yours what to expect when given a diagnosis of gastroschisis.

This is a statewide study. The study will contact approximately 1,600 people in Texas with an invitation to participate. The sponsor is paying for this study to be completed.

PROCEDURES

This study will aim to evaluate the quality of life in children born with gastroschisis.

Caretakers of children born with gastroschisis will be invited to participate in this study.

They will be mailed a general questionnaire and a PedsQL form that asks questions about their child's quality of life. You will be asked to complete and return the questionnaires.

Information from the Texas Department of State Health Services regarding the children of participants who return the questionnaires will be linked with the questionnaire responses and analyzed as part of the study.

If you agree and are able to take part in this study you will be asked to:

- Complete the enclosed questionnaire and PedsQL form and return them to the PI in the provided return envelope.
- By completing and returning the questionnaire, you will be giving your permission to have health information held by the Texas Department of State Health Services linked to your questionnaire responses and analyzed as part of the study.

- The information from the questionnaires and the Texas Department of State Health Services will be linked through the use of a special number so that your name will remain anonymous throughout the entire study.

COMMITMENT

The total amount of time you will take part in this research study is less than 20 minutes. This is the estimated amount of time it will take to fill out the questionnaires. No additional time for follow-up is required from you. The information obtained from the study will be kept until August 2013 to allow for analysis and publication of results.

BENEFITS

The results of this study may help others learn more about the long-term health complications and quality of life of people born with gastroschisis. However, there may be no direct benefit to you for participating in this study.

RISKS AND/OR DISCOMFORTS

This study may include risks that are unknown at this time. The known risks of participating in this study at this time are:

Emotional: You may experience emotional or psychological discomfort when answering questions about your child's experiences related to the gastroschisis.

Confidentiality: Possible risk of breach of confidentiality

Questionnaires: You may get tired when we are asking you questions or you are completing questionnaires. You do not have to answer any questions you do not want to answer.

ALTERNATIVES

If you are willing to have your information used in this study, please return the questionnaires to the Principal Investigator in the provided return envelope. As participation is completely voluntary, all individuals also have the option of not responding to the questionnaires.

STUDY WITHDRAWAL

Your decision to take part is voluntary. You may decide to stop taking part in the study at any time. A decision not to take part or to stop being a part of the research project will not change the services available to you from any physician or service agency.

If you contact the PI with a request to withdraw from the study, none of your information will be included in the results analysis of the study.

COSTS, REIMBURSEMENT AND COMPENSATION

If you decide to take part in this research study, you will not incur any additional costs. You will not be paid for taking part in this study.

CONFIDENTIALITY

You will not be personally identified in any reports or publications that may result from this study. Any personal information about you that is gathered during this study will remain confidential to every extent of the law. A special number (code) will be used to identify you in the study so that you will remain anonymous.

QUESTIONS

If you have questions at any time about this research study, please feel free to contact the PI, Roya Mostafavi at 713-500-5760, as they will be glad to answer your questions. You can contact the study team to discuss problems, voice concerns, obtain information, and offer input in addition to asking questions about the research.

CPHS STATEMENT: This study (HSC-MS-11-0387) has been reviewed by the Committee for the Protection of Human Subjects (CPHS) of the University of Texas Health Science Center at Houston. For any questions about research subject's rights, or to report research-related concerns, call the CPHS at (713) 500-7943. In addition, if you have questions about your rights as a research subject, you may call the Department of State Health Services Institutional Review Board at 512/776-2202 or toll-free at 1-888-777-5037.

Appendix C: Study Questionnaire

Please write your answer in the blank provided. If you do not know an answer, IT IS OKAY TO ESTIMATE:

1. What is the date of birth of the child with gastroschisis? _____
2. What is the current height of the child with gastroschisis? _____ Inches tall
3. What is the current weight of the child with gastroschisis? _____ lbs. _____ oz.
4. How tall is the child's birth mother? _____ Inches tall
5. How much did the child's birth mother weigh before her pregnancy with this child?
_____ lbs
6. Does the child's birth mother have other children, in addition to the child that was born with gastroschisis? Please circle your responses: **Yes** **No**
If yes, how many other children does she have? 1 2 3 4 5 6 7
How old are the children? _____

Please circle your answer to the following questions:

1. What is your relationship to the child that was born with gastroschisis?
 - a. Mother
 - b. Father
 - c. Step-mother
 - d. Step-father
 - e. Grandmother
 - f. Grandfather
 - g. Aunt
 - h. Other: _____
2. From which of the following do you receive any assistance, if any?
 - a. WIC (Women, infants, and children) vouchers
 - b. TANF (Temporary assistance for needy families)
 - c. SNAP (Supplemental Nutrition Assistance Program)
 - d. CHIP (Children's Health Insurance Program)
 - e. School Meals Program
 - f. Low Income Energy Assistance Program
 - g. Child Care Subsidies
 - h. Supplemental security income
 - i. Medicaid
 - j. None

3. What is the annual income of the home?
 - a. Less than \$25,000 per year
 - b. \$25,000 to \$49,999 per year
 - c. \$50,000 to \$74,999 per year
 - d. \$75,000 or more per year
4. At what gestational age was the child with gastroschisis born?
 - a. Full term (\geq 37 weeks)
 - b. Premature
 - c. I do not know
5. The child with gastroschisis was delivered:
 - a. Vaginally
 - b. By C-section
 - c. I do not know
6. The delivery of the child with gastroschisis was:
 - a. Planned or scheduled
 - b. Spontaneous (occurred naturally when the mother went into labor)
 - c. Induced because of medical complications or health reasons
 - d. I do not know
 - e. Other: _____
7. After the child with gastroschisis was born:
 - a. He/she was cared for in the same hospital that he/she was delivered in
 - b. He/she had to be transferred to a different hospital for medical reasons
 - c. I do not know
 - d. Other: _____
8. What kind of surgery did your child have to repair his/her gastroschisis?
 - a. The gastroschisis was repaired in one surgery very soon after birth (primary closure)
 - b. The doctor covered the gastroschisis with an artificial structure and repaired it over the course of a few days (silo placement)
 - c. The doctor covered the gastroschisis with an artificial structure and repaired it several weeks after birth
 - d. I do not know
 - e. Other: _____
9. Has your child needed other surgeries for problems that he or she had since the gastroschisis was first repaired?
 - a. Yes (if yes, please specify type of surgery and age at surgery below)
 - b. No
 - c. I do not remember
 - d. I do not know

If yes, please list additional surgeries (please specify type of surgery and the child's age at each surgery):

Type of surgery: _____ age at surgery: _____

Type of surgery: _____ age at surgery: _____

Type of surgery: _____ age at surgery: _____

10. My child with gastroschisis is:

- a. Much smaller than other children that are the same age
- b. Only slightly smaller than other children that are the same age
- c. The same size as other children that are the same age
- d. Only slightly bigger than other children that are the same age
- e. Much bigger than other children that are the same age

11. My child with gastroschisis has been diagnosed with a learning disability:

- a. Yes
- b. No

If you answered "yes" to question number 11, what learning disability has your child been diagnosed with?

- a. Attention deficit disorder/attention deficit disorder with hyperactivity
- b. Autism/Autism Spectrum such as Aspergers
- c. Developmental delay
- d. Dyslexia
- e. Intellectual disability
- f. Other: _____

12. My child with gastroschisis:

- a. Does not currently attend school
- b. Is currently in pre-school
- c. Is currently in pre-kindergarten
- d. Is currently in kindergarten
- e. Is currently in elementary school
- f. Is currently in middle school
- g. Is currently in high school

13. My child with gastroschisis gets additional help with schoolwork from teachers and tutors:

- a. Never
- b. 1-3 hours per week
- c. 4-6 hours per week
- d. 7-10 hours per week
- e. More than 10 hours per week

14. My child with gastroschisis misses school due to GI problems/discomfort:

- a. 1-2 times per week
- b. 1-2 times per month
- c. Once every 2-3 months
- d. Once every 6 months
- e. Never

15. Please circle any foods that cause your child with gastroschisis to feel uncomfortable:

milk/dairy meat/protein fat/fatty foods chocolate ice cream chips
spicy foods gluten beans raw vegetables sweetener(e.g. sorbitol) sodas
greasy/fried foods
Other: _____

16. My child with gastroschisis needs:

- a. Much more time and care than other children the same age
- b. Slightly more time and care than other children the same age
- c. The same amount of time and care as other children the same age
- d. Slightly less time and care than other children the same age
- e. Much less time and care than other children the same age

Please put an X in the column that best completes the statement about your child with gastroschisis.

	Never	Sometimes	Often	Most of the time	Always
1. My child feels tired:					
2. My child feels self-conscious about the scar from the surgery for the repair of the gastroschisis:					
3. My child is unable to eat certain foods without feeling uncomfortable:					
4. My child feels anxious when there is not a restroom nearby:					
5. My child complains of not being hungry:					

	Rarely	A few times a year	A few times a month	A few times a week	Everyday
1. My child complains of pain in the abdominal area:					
2. My child has problems with reflux:					
3. My child has problems with diarrhea:					
4. My child has problems with constipation:					

My child takes the following medicines:	Never	Daily	Weekly	As Needed
1. Vitamins				
2. Allergy medication				
3. Reflux medication				
4. Diarrhea medication				
5. Nausea/Vomiting medication				
6. Medicine for stomach aches				
7. Medicine for stomach ulcers				
8. Constipation medication				
9. Medicine for gas/bloating				
10. Lactose intolerance medication				
11. Medicine for Ulcerative Colitis/Crohn's /Irritable Bowel				
12. Medicine for ADHD/ADD				
13. Asthma medication				
14. General Pain Relievers (Tylenol, Advil, Ibuprofen)				

How many of the following specialists have your child seen, if any? Indicate all that apply and how often your child sees them.					
	Never Seen	Seen Once	Seen Twice	Seen 3 times	More than 3 times
Audiologist (hearing)					
Cardiologist (heart)					
Developmental Pediatrician (learning problems)					
Ear, nose, and throat doctor					
Gastroenterologist (stomach/digestive system)					
Geneticist					
Nephrologist (kidneys)					
Neurologist (headaches/seizures)					
Pulmonologist (lung/allergies)					
Pediatric Surgeon					
Social Worker					
Physical Therapist					
Occupational Therapist					
Other: _____					

- What do you consider “good quality of life”?

- How would you describe the quality of life of your child with gastroschisis?

- What advice would you give to a parent whose child just received a diagnosis of gastroschisis?

Any other comments for the researchers looking into gastroschisis:

Please fill out the following two pages about Quality of Life and then return both sections of the completed survey in the addressed, stamped envelope to the researchers or contact us at UT Health, Department of Pediatrics, Division of Medical Genetics, Attention Roya Mostafavi, 6431 Fannin Street, MSB 3.142, Houston, Texas, 77030.

Thank you for your participation.

Appendix D: PedsQL 4.0 Scales:

ID# _____

Date: _____

PedsQL
™ Pediatric Quality of Life Inventory

Version 4.0

PARENT REPORT for TODDLERS (ages 2-4)

DIRECTIONS

On the following page is a list of things that might be a problem for **your child**.
Please tell us **how much of a problem** each one has been for **your child**
during the **past ONE month** by circling:

0 if it is **never** a problem

1 if it is **almost never** a problem

2 if it is **sometimes** a problem

3 if it is **often** a problem

4 if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question, please ask for help.

In the past **ONE month**, how much of a **problem** has your child had with ...

PHYSICAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Walking	0	1	2	3	4
2. Running	0	1	2	3	4
3. Participating in active play or	0	1	2	3	4
4. Lifting something heavy	0	1	2	3	4
5. Bathing	0	1	2	3	4
6. Helping to pick up his or her toys	0	1	2	3	4
7. Having hurts or aches	0	1	2	3	4
8. Low energy level	0	1	2	3	4

EMOTIONAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Feeling afraid or scared	0	1	2	3	4
2. Feeling sad or blue	0	1	2	3	4
3. Feeling angry	0	1	2	3	4
4. Trouble sleeping	0	1	2	3	4
5. Worrying	0	1	2	3	4

SOCIAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Playing with other children	0	1	2	3	4
2. Other kids not wanting to play with him or her	0	1	2	3	4
3. Getting teased by other children	0	1	2	3	4
4. Not able to do things that other children his or her age can do	0	1	2	3	4
5. Keeping up when playing with other children	0	1	2	3	4

****Please complete this section if your child attends school or daycare***

SCHOOL FUNCTIONING (<i>problems with...</i>)	Never	Almost Never	Some- times	Often	Almost Always
1. Doing the same school activities as peers	0	1	2	3	4
2. Missing school/daycare because of not feeling well	0	1	2	3	4
3. Missing school/daycare to go to the doctor or Hospital	0	1	2	3	4

ID# _____
Date: _____

TM

PedsQL
Pediatric Quality of Life
Inventory

Version 4.0
PARENT REPORT for YOUNG CHILDREN (ages 5-7)

DIRECTIONS

On the following page is a list of things that might be a problem for **your child**.

Please tell us **how much of a problem** each one has been for **your child**

during the **past ONE month** by circling:

0 if it is **never** a problem

1 if it is **almost never** a problem

2 if it is **sometimes** a problem

3 if it is **often** a problem

4 if it is **almost always** a problem

There are no right or wrong answers.

If you do not understand a question, please ask for help.

In the past **ONE month**, how much of a **problem** has your child had with ...

PHYSICAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Walking more than one block	0	1	2	3	4
2. Running	0	1	2	3	4
3. Participating in sports activity or exercise	0	1	2	3	4
4. Lifting something heavy	0	1	2	3	4
5. Taking a bath or shower by him or herself	0	1	2	3	4
6. Doing chores, like picking up his or her toys	0	1	2	3	4
7. Having hurts or aches	0	1	2	3	4
8. Low energy level	0	1	2	3	4

EMOTIONAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Feeling afraid or scared	0	1	2	3	4
2. Feeling sad or blue	0	1	2	3	4
3. Feeling angry	0	1	2	3	4
4. Trouble sleeping	0	1	2	3	4
5. Worrying about what will happen to him or her	0	1	2	3	4

SOCIAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Getting along with other children	0	1	2	3	4
2. Other kids not wanting to be his or her friend	0	1	2	3	4
3. Getting teased by other children	0	1	2	3	4
4. Not able to do things that other children his or her age can do	0	1	2	3	4
5. Keeping up when playing with other children	0	1	2	3	4

SCHOOL FUNCTIONING (<i>problems with...</i>)	Never	Almost Never	Some- times	Often	Almost Always
1. Paying attention in class	0	1	2	3	4
2. Forgetting things	0	1	2	3	4
3. Keeping up with school activities	0	1	2	3	4
4. Missing school because of not feeling well	0	1	2	3	4
5. Missing school to go to the doctor or hospital	0	1	2	3	4

ID# _____

Date: _____

TM
PedsQL
Pediatric Quality of Life
Inventory

Version 4.0

PARENT REPORT for CHILDREN (ages 8-12)

DIRECTIONS

On the following page is a list of things that might be a problem for **your child**.

Please tell us **how much of a problem** each one has been for **your child** during the **past ONE month** by circling:

0 if it is **never** a problem

1 if it is **almost never** a problem

2 if it is **sometimes** a problem

3 if it is **often** a problem

4 if it is **almost always** a problem

There are no right or wrong answers.

If you do not understand a question, please ask for help.

In the past **ONE month**, how much of a **problem** has your child had with ...

PHYSICAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Walking more than one block	0	1	2	3	4
2. Running	0	1	2	3	4
3. Participating in sports activity or exercise	0	1	2	3	4
4. Lifting something heavy	0	1	2	3	4
5. Taking a bath or shower by him or herself	0	1	2	3	4
6. Doing chores around the house	0	1	2	3	4
7. Having hurts or aches	0	1	2	3	4
8. Low energy level	0	1	2	3	4

EMOTIONAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Feeling afraid or scared	0	1	2	3	4
2. Feeling sad or blue	0	1	2	3	4
3. Feeling angry	0	1	2	3	4
4. Trouble sleeping	0	1	2	3	4
5. Worrying about what will happen to him or her	0	1	2	3	4

SOCIAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Getting along with other children	0	1	2	3	4
2. Other kids not wanting to be his or her friend	0	1	2	3	4
3. Getting teased by other children	0	1	2	3	4
4. Not able to do things that other children his or her age can do	0	1	2	3	4
5. Keeping up when playing with other children	0	1	2	3	4

SCHOOL FUNCTIONING (<i>problems with...</i>)	Never	Almost Never	Some- times	Often	Almost Always
1. Paying attention in class	0	1	2	3	4
2. Forgetting things	0	1	2	3	4
3. Keeping up with schoolwork	0	1	2	3	4
4. Missing school because of not feeling well	0	1	2	3	4
5. Missing school to go to the doctor or hospital	0	1	2	3	4

Appendix E: Spanish Translations:

Invitación a participar en una encuesta sobre la salud y la calidad de vida de los niños con gastrosquisis

Estimado padre, madre o tutor:

Se le invita a ayudar en una investigación sobre los efectos a largo plazo de la gastrosquisis en la calidad de vida y la salud de su hijo. El propósito de esta investigación es entender mejor las experiencias de los niños nacidos con gastrosquisis para que los profesionales de salud puedan explicarle mejor a las familias como la suya qué esperar.

La encuesta es anónima y opcional. Los resultados se usarán sólo para fines de investigación. Debe tomarle menos de 20 minutos para rellenar la encuesta. Hemos adjuntado las versiones tanto en español como en inglés y puede usted usar el idioma que prefiera.

SI USTED ACEPTA PARTICIPAR, SIGA ESTAS INDICACIONES:

- La persona que cuida al niño con gastrosquisis debe rellenar todas las partes de la encuesta.
- Conteste lo más completa y sinceramente que pueda.
- Puede elegir saltar cualquier pregunta que no quiera contestar y puede dejar de hacer la encuesta en cualquier momento.
- Al terminar, ponga los formularios rellenos en el sobre de respuesta provisto y mándelos por correo postal lo más pronto posible.

INFORMACIÓN IMPORTANTE SOBRE SU PRIVACIDAD:

- Todas las respuestas son confidenciales.
- Se resumirán y presentarán los datos como parte de un proyecto de tesis de la Facultad de Ciencias Biomédicas de la Universidad de Texas en Houston.
- Aunque los resultados del estudio podrían ser presentados para su publicación, su nombre no se vinculará con sus respuestas y su nombre no aparecerá en ningún informe ni publicación.
- No se le pagará por hacer la encuesta.
- Si elige no participar o cambia de parecer después de iniciar su participación, no perderá ninguna de sus prestaciones.
- El Registro de Defectos de Nacimiento de Texas es parte del Departamento Estatal de Servicios de Salud de Texas que reúne información sobre los bebés con defectos de nacimiento para vigilar los índices de los defectos de nacimiento, buscar las causas de los defectos de nacimiento, mejorar la salud pública e informar a las familias sobre los servicios disponibles.

Los resultados del estudio podrían ayudar a otras personas a informarse más sobre las complicaciones de salud a largo plazo y la calidad de vida de la gente con gastrosquisis. Sin embargo, podría no haber ningún beneficio directo para usted al participar en el estudio. La encuesta ha sido aprobada por las juntas de revisión institucional tanto del Departamento

Estatad de Servicios de Salud de Texas como del Centro de Ciencia de la Salud de la Universidad de Texas en Houston.

Si tiene alguna pregunta o duda, contacte a Roya Mostafavi, BS en Roya.M.Mostafavi@uth.tmc.edu o llamando al 713-500-5760. Además, si tiene preguntas sobre sus derechos como sujeto de investigación, puede llamar a la Junta de Revisión Institucional del Departamento Estatal de Servicios de Salud de Texas, al teléfono 512-776-2202, o gratis al 1-888-777-5037.

Muchas gracias por ayudarnos con esta importante investigación.

Atentamente,

Roya Mostafavi, BS UT Health Pasante de asesoramiento genético Investigadora principal	Claire Singletary, MS CGC UT Health Directora, Programa de Asesoramiento Genético Presidente del comité	Karen Moffitt, MPH División de Epidemiología y Vigilancia de Defectos de Nacimiento de Texas Depto. Estatal de Servicios de Salud de Texas
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FORMULARIO DE CONSENTIMIENTO INFORMADO PARA PARTICIPAR EN UNA INVESTIGACIÓN

Análisis de resultados a largo plazo de morbilidad y calidad de vida de pacientes que nacieron con gastrosquisis

HSC-MS-11-0387
TDSHS IRB #11-065

INVITACIÓN A PARTICIPAR

Lo invitamos a participar en un proyecto de investigación conocido como *Análisis de resultados a largo plazo de morbilidad y calidad de vida de pacientes que nacieron con gastrosquisis*, realizado por Roya Mostafavi, BS, del Centro de Ciencia de la Salud de la Universidad de Texas. En lo que respecta a este proyecto de investigación, se le conocerá como la Investigadora principal, o PI.

Su decisión de participar es voluntaria. Puede negarse a participar o elegir dejar de participar en cualquier momento. La decisión de no participar o dejar de participar en el proyecto de investigación no cambiará los servicios que están a su disposición.

Puede negarse a contestar cualquiera de las preguntas que le hagan en cualquiera de los formularios. Este proyecto de investigación ha sido revisado por el Comité para la Protección de los Sujetos Humanos (o CPHS) del Centro de Ciencia de la Salud de la Universidad de Texas en Houston como HSC-MS-11-0387. El estudio también ha sido revisado por el Departamento Estatal de Servicios de Salud de Texas como IRB #11-065.

PROPÓSITO

El propósito de este estudio de investigación es entender mejor las experiencias de los niños nacidos con gastrosquisis para que los profesionales de salud puedan explicarle mejor a las familias como la suya qué esperar al recibir un diagnóstico de gastrosquisis.

Se trata de un estudio estatal. En el estudio contactaremos a aproximadamente 1,600 personas de Texas para invitarlos a participar. El patrocinador está pagando para que se finalice este estudio.

PROCEDIMIENTOS

Este estudio tiene como objetivo evaluar la calidad de vida de los niños que nacieron con gastrosquisis. Se invitará a cuidadores de niños que nacieron con gastrosquisis a participar en este estudio. Les mandarán un cuestionario general y un formulario PedsQL, en el cual habrá preguntas sobre la calidad de vida de su hijo. Le pedirán que rellene y devuelva los cuestionarios. Se relacionará información del Departamento Estatal de Servicios de Salud de Texas sobre los hijos de los participantes que devuelvan los cuestionarios con las respuestas de dichos cuestionarios y se analizará como parte del estudio.

Si usted acepta y puede participar en este estudio, le pedirán que:

- Rellene el cuestionario y formulario PedsQL adjuntos y los devuelva a la PI en el sobre de respuesta provisto.
- Al rellenar y devolver el cuestionario, estará dando su permiso para que la información de salud que retiene el Departamento Estatal de Servicios de Salud de Texas se relacione con sus respuestas del cuestionario y se analice como parte del estudio.

- Se relacionará la información de los cuestionarios con la del Departamento Estatal de Servicios de Salud de Texas mediante el uso de un número especial para que su nombre siga siendo anónimo durante todo el estudio.

COMPROMISO

El tiempo total que usted participará en este estudio de investigación será menos de 20 minutos. Ese es el tiempo aproximado que tomará para rellenar los cuestionarios. No se requerirá más de su tiempo para dar seguimiento. La información obtenida del estudio se guardará hasta agosto de 2013 para permitir el análisis y la publicación de los resultados.

BENEFICIOS

Los resultados del estudio podrían ayudar a otras personas a informarse más sobre las complicaciones de salud a largo plazo y la calidad de vida de las personas que nacieron con gastrosquisis. Sin embargo, podría no haber ningún beneficio directo para usted al participar en el estudio.

RIESGOS O MOLESTIAS

Este estudio podría incluir riesgos que se desconocen en este momento. En este momento los riesgos que se conocen de participar en este estudio son:

Emocionales: Podría experimentar molestia emocional o psicológica al contestar preguntas sobre las experiencias de su hijo relacionadas con la gastrosquisis.

Confidencialidad: Posible riesgo de violación de la confidencialidad

Cuestionarios: Podría cansarse al nosotros hacerle preguntas o al usted rellenar los cuestionarios. No tiene que contestar ninguna pregunta que no quiera contestar.

ALTERNATIVAS

Si está dispuesto a que se use su información en este estudio, devuelva los cuestionarios a la Investigadora principal en el sobre de respuesta provisto. Ya que la participación es completamente voluntaria, todos los individuos también tienen la opción de no responder a los cuestionarios.

RETIRADA DEL ESTUDIO

Su decisión de participar es voluntaria. Puede decidir dejar de participar en el estudio en cualquier momento. La decisión de no participar o dejar de participar en el proyecto de investigación no cambiará los servicios que están a su disposición de cualquier médico o agencia de servicio.

Si usted se comunica con la PI con una petición de retirada del estudio, no se incluirá ninguna de la información suya en el análisis de los resultados del estudio.

GASTOS, REEMBOLSOS Y REMUNERACIÓN

Si decide participar en este estudio de investigación, no tendrá ningún gasto adicional. No le pagarán por participar en este estudio.

CONFIDENCIALIDAD

No se le identificará personalmente en ningún informe ni publicación que pudiera resultar de este estudio. Se mantendrá la confidencialidad de cualquier información personal sobre usted que se reúna durante este estudio según la ley en toda su extensión. Se usará un número especial (código) para identificarlo en el estudio para que usted siga siendo anónimo.

PREGUNTAS

Si en cualquier momento tiene preguntas sobre este estudio de investigación, no dude en llamar a la PI, Roya Mostafavi, al 713-500-5760, ya que con gusto contestará sus preguntas. Usted puede comunicarse con el equipo del estudio para hablar de problemas, expresar preocupaciones, obtener información y ofrecer su opinión, además de hacer preguntas sobre la investigación.

DECLARACIÓN DEL CPHS: Este estudio (HSC-MS-11-0387) ha sido revisado por el Comité para la Protección de los Sujetos Humanos (CPHS) del Centro de Ciencia de la Salud de la Universidad de Texas en Houston. Si tiene preguntas sobre los derechos de los sujetos de investigación o para informar de dudas relacionadas con la investigación, llame al CPHS al (713) 500-7943. Además, si tiene preguntas sobre sus derechos como sujeto de investigación, puede llamar a la Junta de Revisión Institucional del Departamento Estatal de Servicios de Salud de Texas, al teléfono 512-776-2202, o gratis al 1-888-777-5037.

Escriba su respuesta en el espacio en blanco provisto. Si no sabe una respuesta, PUEDE HACER UN CÁLCULO APROXIMADO:

1. ¿Cuál es la fecha de nacimiento del niño con gastrosquisis? _____
2. ¿Cuál es la estatura actual del niño con gastrosquisis? _____ Pulgadas de alto
3. ¿Cuál es el peso actual del niño con gastrosquisis? _____ Lb _____ oz
4. ¿Cuánto mide la madre biológica del niño?
_____ Pulgadas de alto
5. ¿Cuánto pesaba la madre biológica del niño antes de estar embarazada con dicho niño? _____ Lb
6. ¿Tiene otros hijos la madre biológica además del niño que nació con gastrosquisis?

Encierre sus respuestas en un círculo:

Sí

No

Si “sí”, ¿cuántos hijos más tiene? 1 2 3 4 5 6 7

¿Qué edades tienen los niños?

Encierre en un círculo su respuesta a las siguientes preguntas:

1. ¿Cuál es su parentesco con el niño que nació con gastrosquisis?
 - a. Madre
 - b. Padre
 - c. Madrastra
 - d. Padrastro
 - e. Abuela
 - f. Abuelo
 - g. Tía
 - h. Otro: _____
2. ¿De cuáles de los siguientes recibe asistencia, de recibirla?
 - a. Vales del WIC (Programa para Mujeres, Bebés y Niños)
 - b. TANF (Asistencia Temporal para Familias Necesitadas)
 - c. SNAP (Programa de Asistencia de Nutrición Suplemental)
 - d. CHIP (Programa de Seguro Médico Infantil)
 - e. Programa de Alimentos Escolares
 - f. Programa de Asistencia con los Cobros de Luz para Personas de Escasos Ingresos
 - g. Subsidios para el cuidado infantil
 - h. Ingresos de Seguridad Suplemental
 - i. Medicaid
 - j. Ninguno

3. ¿Cuáles son los ingresos anuales de la casa?
 - a. Menos de \$25,000 al año
 - b. \$25,000 a \$49,999 al año
 - c. \$50,000 a \$74,999 al año
 - d. \$75,000 al año o más
4. ¿A qué edad gestacional nació el niño con gastrosquisis?
 - a. A término (nació a las 37 semanas o después)
 - b. Prematuro
 - c. No sé
5. El niño con gastrosquisis nació por parto:
 - a. vaginal
 - b. por cesárea
 - c. no sé
6. El parto del niño con gastrosquisis fue:
 - a. planeado o programado
 - b. espontáneo (ocurrió naturalmente cuando la madre empezó el trabajo de parto)
 - c. inducido debido a complicaciones médicas o razones de salud
 - d. no sé
 - e. otro: _____
7. Después de nacer, el niño con gastrosquisis:
 - a. fue atendido en el mismo hospital donde nació
 - b. tuvo que ser transferido a otro hospital por razones médicas
 - c. no sé
 - d. otro: _____
8. ¿Qué tipo de cirugía le hicieron a su hijo para reparar la gastrosquisis?
 - a. La gastrosquisis fue reparada en una sola cirugía muy poco después del nacimiento (cierre primario)
 - b. El doctor cubrió la gastrosquisis con una estructura artificial y la reparó en el transcurso de varios días (colocación de silo)
 - c. El doctor cubrió la gastrosquisis con una estructura artificial y la reparó varias semanas después del nacimiento
 - d. No sé
 - e. Otro: _____

9. ¿Ha necesitado otras cirugías su hijo por problemas que tuvo él o ella desde que la gastrosquisis se reparó por primera vez?
- a. Sí (si contestó que “sí”, especifique abajo el tipo de cirugía y qué edad tenía al hacerse la cirugía)
 - b. No
 - c. No recuerdo
 - d. No sé

Sí “sí”, liste las cirugías adicionales (especifique el tipo de cirugía y qué edad tenía el niño al hacerse la cirugía):

Tipo de cirugía: _____ edad al hacerse la cirugía: _____

Tipo de cirugía: _____ edad al hacerse la cirugía: _____

Tipo de cirugía: _____ edad al hacerse la cirugía: _____

10. Mi hijo con gastrosquisis es:

- a. mucho más pequeño que otros niños de la misma edad
- b. sólo un poco más pequeño que otros niños de la misma edad
- c. del mismo tamaño que otros niños de la misma edad
- d. sólo un poco más grande que otros niños de la misma edad
- e. mucho más grande que otros niños de la misma edad

11. A mi hijo con gastrosquisis se le ha diagnosticado una discapacidad de aprendizaje:

- a. sí
- b. no

Si contestó que “sí” a la pregunta número 11, ¿qué discapacidad de aprendizaje le han diagnosticado a su hijo?

- a. Trastorno por déficit de atención (ADD) o trastorno por déficit de atención con hiperactividad (ADHD)
- b. Autismo o espectro de autismo como el síndrome de Asperger
- c. Retraso de desarrollo
- d. Dislexia
- e. Discapacidad intelectual
- f. Otra: _____

12. Mi hijo con gastrosquisis:

- a. actualmente no asiste a la escuela
- b. actualmente está en rescolar
- c. actualmente está en prekínder
- d. actualmente está en kínder
- e. actualmente está en escuela primaria
- f. actualmente está en middle school
- g. actualmente está en high school

13. Mi hijo con gastrosquiasis recibe ayuda adicional de los maestros y los tutores con el trabajo escolar:

- a. nunca
- b. 1-3 horas a la semana
- c. 4-6 horas a la semana
- d. 7-10 horas a la semana
- e. más de 10 horas a la semana

14. Mi hijo con gastrosquiasis falta a la escuela debido a problemas o molestias gastrointestinales (GI):

- a. 1-2 veces a la semana
- b. 1-2 veces al mes
- c. una vez cada 2-3 meses
- d. una vez cada 6 meses
- e. nunca

15. Encierre en un círculo los alimentos que le causan molestia a su hijo con gastrosquiasis :

leche o alimentos lácteos carne o proteína grasa o alimentos grasos
chocolate helado totopos alimentos picantes gluten frijoles
verduras crudas endulzante (p. ej. Sorbitol) refrescos alimentos grasos
o fritos
otro: _____

16. Mi hijo con gastrosquiasis necesita:

- a. mucho más tiempo y cuidado que otros niños de la misma edad
- b. un poco más tiempo y cuidado que otros niños de la misma edad
- c. el mismo tiempo y cuidado que otros niños de la misma edad
- d. un poco menos tiempo y cuidado que otros niños de la misma edad
- e. mucho menos tiempo y cuidado que otros niños de la misma edad

Ponga una “X” en la columna que mejor completa la declaración sobre su hijo con gastrosquisis.

	Nunca	A veces	A menudo	La mayoría del tiempo	Siempre
1. Mi hijo se siente cansado:					
2. Mi hijo se siente acomplexado por la cicatriz de la cirugía para reparar la gastrosquisis:					
3. Mi hijo no puede comer ciertos alimentos sin sentir molestia:					
4. Mi hijo se siente ansioso si no está cerca de un baño:					
5. Mi hijo se queja de no tener hambre:					

	Rara vez	Varias veces al año	Varias veces al mes	Varias veces a la semana	Todos los días
1. Mi hijo se queja de dolor del área abdominal:					
2. Mi hijo tiene problemas de reflujo:					
3. Mi hijo tiene problemas de diarrea:					
4. Mi hijo tiene problemas de estreñimiento:					

Mi hijo toma las siguientes medicinas:	Nunca	Cada día	Cada semana	De ser necesario
1. Vitaminas				
2. Medicina para las alergias				
3. Medicina para el reflujo				
4. Medicina para la diarrea				
5. Medicina para la náusea o el vómito				
6. Medicina para los dolores estomacales				
7. Medicina para las úlceras estomacales				
8. Medicina para el estreñimiento				
9. Medicina para el gas o la hinchazón				
10. Medicina para la intolerancia a la lactosa				
11. Medicina para la colitis ulcerativa, la enfermedad de Crohn o el síndrome del intestino irritable				
12. Medicina para el ADHD o el ADD				
13. Medicina para el asma				
14. Medicinas para el dolor general (Tylenol, Advil o ibuprofeno)				

¿Cuántos de los siguientes especialistas han atendido a su hijo, de haber alguno? Indique todos los que correspondan y con qué frecuencia atienden a su hijo.					
	Nunca lo ha atendido	Lo ha atendido una vez	Lo ha atendido dos veces	Lo ha atendido 3 veces	Más de 3 veces
Audiólogo (audición)					
Cardiólogo (corazón)					
Pediatra del desarrollo (problemas de aprendizaje)					
Doctor del oído, nariz, y garganta					
Gastroenterólogo (estómago o aparato digestivo)					
Genetista					
Nefrólogo (riñones)					
Neurólogo (dolores de cabeza o ataques)					
Neumólogo (pulmones o alergias)					
Cirujano pediátrico					
Trabajador social					
Fisioterapeuta					
Terapeuta ocupacional					
Otro: _____					

4. ¿Qué considera usted que es una “buena calidad de vida”?

2. ¿Cómo describiría la calidad de vida de su hijo con gastrosquisis?

5. ¿Qué consejos le daría a un padre o madre a cuyo hijo acaban de diagnosticarle gastrosquisis?

Cualquier otro comentario para los investigadores que estudian la gastrosquisis:

Rellene las siguientes dos páginas sobre la calidad de vida y devuelva ambas secciones de la encuesta rellena en el sobre con franqueo pagado con los datos del remitente a los investigadores o contáctenos en UT Health, Department of Pediatrics, Division of Medical Genetics, Attention Roya Mostafavi, 6431 Fannin Street, MSB 3.142, Houston, Texas, 77030.

Gracias por su participación.

ID# _____
Fecha: _____

PedsQLTM

Inventario Sobre Calidad de Vida Pediátrica

Versión 4.0

REPORTE de PADRES para NIÑOS (edades 2-4)

INSTRUCCIONES

En la página siguiente hay una lista de cosas que pudieran ser un problema para **su hijo(a)**. Por favor díganos **cuánto problema** ha sido esto para **su hijo(a)** durante el **mes pasado (UN mes)**. Por favor circule su respuesta:

- 0 si **nunca** es un problema
- 1 si **casi nunca** es un problema
- 2 si **algunas veces** es un problema
- 3 si **a menudo** es un problema
- 4 si **casi siempre** es un problema

No hay respuestas correctas o incorrectas.
Si Ud. no entiende una pregunta, por favor pida ayuda.

*En el mes pasado (Un mes), cuánto **problema** ha tenido su hijo(a) con ...*

PedsQL 4.0 – Parent (2-4)
Spanish/Broadcast (02/01)

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FUNCIONAMIENTO FÍSICO (problemas con...)	Nunca	Casi nunca	Algunas Veces	A Menudo	Casi Siempre
1. Caminado	0	1	2	3	4
2. Corriendo	0	1	2	3	4
3. Participando en juegos activos o ejercicios	0	1	2	3	4
4. Levantando algo pesado	0	1	2	3	4
5. Bañándose	0	1	2	3	4
6. Ayudando a recoger sus juguetes	0	1	2	3	4
7. Teniendo dolores o molestias	0	1	2	3	4
8. Poca energía	0	1	2	3	4

FUNCIONAMIENTO EMOCIONAL (problemas con...)	Nunca	Casi Nunca	Algunas Veces	A Menudo	Casi Siempre
1. Sintiendo asustado o con miedo	0	1	2	3	4
2. Sintiendo triste o decaído	0	1	2	3	4
3. Sintiendo enojado	0	1	2	3	4
4. Dificultades para dormir	0	1	2	3	4
5. Preocupándose	0	1	2	3	4

FUNCIONAMIENTO SOCIAL (problemas con...)	Nunca	Casi Nunca	Algunas Veces	A Menudo	Casi Siempre
1. Jugando con otros niños	0	1	2	3	4
2. Otros niños no queriendo jugar con él o ella	0	1	2	3	4
3. Otros niños burlándose de él o ella	0	1	2	3	4
4. No pudiendo hacer cosas que otros niños de su edad pueden hacer	0	1	2	3	4
5. Pudiendo mantenerse al igual con otros niños cuando juega	0	1	2	3	4

Por favor complete esta sección si su niño(a) asiste a la escuela o a la guardería

FUNCIONAMIENTO ESCOLAR (problemas con...)	Nunca	Casi Nunca	Algunas Veces	A Menudo	Casi Siempre
1. Haciendo las mismas actividades escolares que sus compañeros	0	1	2	3	4
2. Faltando a la escuela/guardería porque no se siente bien	0	1	2	3	4
3. Faltando a la escuela/guardería para ir al doctor o al hospital	0	1	2	3	4

ID# _____
Fecha: _____

PedsQLTM

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Versión 4.0

REPORTE de PADRES para NIÑOS (edades 5-7)

INSTRUCCIONES

En la página siguiente hay una lista de cosas que pudieran ser un problema para su hijo(a). Por favor díganos cuánto problema ha sido esto para su hijo(a) durante el mes pasado (UN mes). Por favor circule su respuesta:

- 0 si nunca es un problema
- 1 si casi nunca es un problema
- 2 si algunas veces es un problema
- 3 si a menudo es un problema
- 4 si casi siempre es un problema

No hay respuestas correctas o incorrectas.
Si Ud. no entiende una pregunta, por favor pida ayuda.

En el mes pasado (**UN mes**), cuánto **problema** ha tenido su hijo(a) con ...

FUNCIONAMIENTO FÍSICO (problemas con...)	Nunca	Casi nunca	Algunas Veces	A Menudo	Casi Siempre
1. Caminado más de una cuadra	0	1	2	3	4
2. Corriendo	0	1	2	3	4
3. Participando en actividades deportivas o ejercicios	0	1	2	3	4
4. Levantando algo pesado	0	1	2	3	4
5. Tomando una ducha o tina por sí mismo(a)	0	1	2	3	4
6. Haciendo quehaceres, como recoger sus juguetes	0	1	2	3	4
7. Teniendo dolores o molestias	0	1	2	3	4
8. Poca energía	0	1	2	3	4

FUNCIONAMIENTO EMOCIONAL (problemas con...)	Nunca	Casi Nunca	Algunas Veces	A Menudo	Casi Siempre
1. Sintiendo asustado o con miedo	0	1	2	3	4
2. Sintiendo triste o decaído	0	1	2	3	4
3. Sintiendo enojado	0	1	2	3	4
4. Dificultades para dormir	0	1	2	3	4
5. Preocupándose por lo que le vaya a pasar	0	1	2	3	4

FUNCIONAMIENTO SOCIAL (problemas con...)	Nunca	Casi Nunca	Algunas Veces	A Menudo	Casi Siempre
1. Levándose bien con otros niños	0	1	2	3	4
2. Otros niños no queriendo ser amigos de él o ella	0	1	2	3	4
3. Otros niños burlándose de él o ella	0	1	2	3	4
4. No pudiendo hacer cosas que otros niños de su edad pueden hacer	0	1	2	3	4
5. Pudiendo mantenerse al igual con otros niños cuando juega	0	1	2	3	4

FUNCIONAMIENTO ESCOLAR (problemas con...)	Nunca	Casi Nunca	Algunas Veces	A Menudo	Casi Siempre
1. Poniendo atención en clase	0	1	2	3	4
2. Olvidando cosas	0	1	2	3	4
3. Manteniéndose al día con actividades escolares	0	1	2	3	4
4. Faltando a la escuela porque no se siente bien	0	1	2	3	4
5. Faltando a la escuela para ir al doctor o al hospital	0	1	2	3	4

ID#	_____
Fecha:	_____

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Versión 4.0

REPORTE de PADRES para NIÑOS (edades 8-12)

INSTRUCCIONES

En la página siguiente hay una lista de cosas que pudieran ser un problema para su hijo(a). Por favor díganos **cuánto problema** ha sido esto para su hijo(a) durante el mes pasado (**UN mes**). Por favor circule su respuesta:

- 0 si **nunca** es un problema
- 1 si **casi nunca** es un problema
- 2 si **algunas veces** es un problema
- 3 si **a menudo** es un problema
- 4 si **casi siempre** es un problema

No hay respuestas correctas o incorrectas.

Si Ud. no entiende una pregunta, por favor pida ayuda.

*En el mes pasado (**UN mes**), cuánto **problema** ha tenido su hijo(a) con ...*

FUNCIONAMIENTO FÍSICO (<i>problemas con...</i>)	Nunca	Casi nunca	Algunas Veces	A Menudo	Casi Siempre
1. Caminando más de una cuadra	0	1	2	3	4
2. Corriendo	0	1	2	3	4
3. Participando en actividades deportivas o ejercicios	0	1	2	3	4
4. Levantando algo pesado	0	1	2	3	4
5. Tomando una ducha o tina por sí mismo(a)	0	1	2	3	4
6. Haciendo quehaceres en la casa	0	1	2	3	4
7. Teniendo dolores o molestias	0	1	2	3	4
8. Poca energía	0	1	2	3	4

FUNCIONAMIENTO EMOCIONAL (<i>problemas con...</i>)	Nunca	Casi Nunca	Algunas Veces	A Menudo	Casi Siempre
1. Sintiendo asustado o con miedo	0	1	2	3	4
2. Sintiendo triste o decaído	0	1	2	3	4
3. Sintiendo enojado	0	1	2	3	4
4. Dificultades para dormir	0	1	2	3	4
5. Preocupándose por lo que le vaya a pasar	0	1	2	3	4

FUNCIONAMIENTO SOCIAL (<i>problemas con...</i>)	Nunca	Casi Nunca	Algunas Veces	A Menudo	Casi Siempre
1. Llevándose bien con otros niños	0	1	2	3	4
2. Otros niños no queriendo ser amigos de él o ella	0	1	2	3	4
3. Otros niños burlándose de él o ella	0	1	2	3	4
4. No pudiendo hacer cosas que otros niños de su edad pueden hacer	0	1	2	3	4
5. Pudiendo mantenerse al igual con otros niños cuando juega	0	1	2	3	4

FUNCIONAMIENTO ESCOLAR (<i>problemas con...</i>)	Nunca	Casi Nunca	Algunas Veces	A Menudo	Casi Siempre
1. Poniendo atención en clase	0	1	2	3	4
2. Olvidando cosas	0	1	2	3	4
3. Manteniéndose al día con actividades escolares	0	1	2	3	4
4. Faltando a la escuela porque no se siente bien	0	1	2	3	4
5. Faltando a la escuela para ir al doctor o al hospital	0	1	2	3	4

Appendix F: Free Response Questions:

1. What do you consider “good quality of life”?

- “Good overall health.”
- “Good, clean, healthy living with family.”
- “Healthy, smiles, activity.”
- “Enjoying good health, happy family life.”
- "He is a healthy child with a beautiful scar. That is all."
- “Being a boy like any other, only with a scar on his stomach, but that he views himself and acts like any other boy his age.”
- “Being able to function as best as same age peers in cognitive and physical abilities. He is a baseball star, stronger than most kids twice his age and an avid writer.”
- “Being able to take care of yourself and function in a normal capacity of life.”
- “Living healthy, cheerful, happy. A good quality of life produces ‘happiness’ and that is what it is. Thanks to God.”
- “Good health, equivalent to peers, and happy.”
- “The same as other children his age and household income.”

- “To me, ‘good quality of life’ is when a person is able to function normally in society, without physical limitations restricting them from performing their daily activities and enjoying the things they love.”
- “Being able to do the same things as kids his age without restrictions.”
- “Able to function and participate without much physical or mental distractions.”
- “Being happy and healthy.”
- “Live your life as normal as possible and in good health.”
- “Healthy”
- “that we are healthy”
- “A good diet and regulated care”
- “Life lived without health and finance boundaries”
- “The ability to function and contribute on par with those around you”
- “Good quality of life is being able to wake up every morning pain free and happy without the fear of sickness and disease controlling your life.”
- “My son has a "good quality of life. He is 7 years old. Recovered from surgery at 6 weeks. Now is in accelerated reading and about to be tested for Gifted/Talented.”
- "In which the child grows normally, without any complications"
- “Living healthy and happy, in a good family environment full of love and care."

- “A healthy, full of energy, happy, full of joy, full of confidence child, and smart child with a huge heart! One who forgives. To be able to travel around the world.”
- “Physical health, family/friends, workplace, home”
- “Healthy, happy, active, and not knowing what he went through.”
- “Very little pain, ability to communicate, and having the emotional capacity to feel joy or happiness.”
- “Normal playing kid gets to do all kid activities”
- “being able to do other things do the same age”
- “A good quality of life includes being able to do what you want to do socially, professionally, and mentally, and having the monetary wherewithal to support yourself.”
- “Able to complete everyday tasks with little to no aid, emotionally and socially involved in the world around you; being around good people.”
- “Lots of learning, reading, loving, caring in the family. Needs to know that I would do anything for them.”
- “Being healthy-physically, mentally, emotionally. Loving and supportive family.”
- “Happy, healthy child in loving, stable home environment that is able to participate with their peers.”
- “healthy and happy”

- “My son has a good quality of life but the first year of his life he didn't; he just spent it in the hospital, he had many problems.”
- “No problems physically and emotionally with the child. Good healthy and happy lifestyle.”
- “healthy and happy”
- “Good health habits, going to the doctor's office for regular checkups, and providing nutritional foods.”
- “If the child is healthy, happy and able to perform most or all tasks needed to play, socialize, or function. I believe that that is a good quality of life.”
- “Having a life where they don't have to be hooked up to monitors and machines all their life.”
- “A normal kid life, loving family, stable household, health, and discipline.”
- “Having my child being healthy”
- “Full of family, happiness, and good times.”

2. How would you describe the quality of life of your child with gastroschisis?

- “Fair. He/we have struggled with learning disabilities, vision, low self-esteem and a few others.”
- “He's doing great.”
- "good"

- "She is very good in school, holds an A average. She is happy in her own skin and loves others. She has a good quality of life."
- "Very good. Some problems focusing and shows attitude (pretty normal attitude for preteen)."
- "Good. Normal for his age."
- "He can accomplish anything just like any other kid."
- "His quality of life has not been altered by gastroschisis. He is above level on most cognitive abilities as well as physical."
- "Very active, no restriction whatsoever."
- "Excellent!!"
- "good"
- "Very good"
- "She does have a few mild physical limitations, but they are not bothersome enough to keep her from doing activities that other kids her age do. Her quality of life is good."
- "He has the best quality of life, he plays sports just like a normal child and doesn't have any problems from this."
- "great"

- "Good. To my knowledge she is almost always happy. She has a ton of friends. She gets very embarrassed though about the two scars from her surgeries. We are considering plastic surgery for the scars when she gets older."
- "The same as other children his age. He does the same things as his siblings and, in general, is a very healthy boy."
- "He is very healthy, smart, and active."
- "His quality is great. I believe he is great. The only problem he has had is that he can't get spicy food."
- "good"
- "My child is happy, healthy, and spoiled. For a child, his quality of life is wonderful and limitless."
- "Good"
- "Her quality of life is not terrible, however it could be better. She has abdominal pain and discomfort every day, but she is learning how to manage and deal with it. She is a very happy child."
- "[He] shows no signs of differences with his birth defect. He smiles, plays, and cares nothing about scar yet."
- "Good, normal growth, and without any complications related to the gastroschisis he suffered from at birth (he only has language problems)."

- “Good, my daughter is a girl like any other, except that she doesn't have a belly button like other children.”
- "Excellent, he is a very energetic kid, who lives a normal lifestyle. He knows he has had surgery when born and he is proud of it. He is in taekwondo, basketball, Chinese classes, aside from kindergarten school. He eats everything I put on the table. He is as any 5 year old."
- "My son is a very energetic athletic child very heart-warming with a good sense of humor. He likes telling jokes. He loves his family."
- "Healthy and happy. Very active, normal, little 5 year old boy. Doesn't know unless we told him what he went through."
- "She has been mostly unaffected other than the surgery at birth and three weeks in the NICU. As an infant she actually had less digestive problems than my older child. She is exceptionally intelligent and a very happy content child."
- "Limited to some activities. A little worried someone may hit his belly too hard when playing."
- "My child's life and quality is not any different than other kids her age. The only noticeable thing is her scar on her abdomen."
- "Good quality. Normal to other children after the first year."
- "My child has a very good quality of life filled with many people who love and care for him."

- "My son is blessed in the fact that his gastroschisis has had a minimal impact on his development. Unless he shows you his scar (which he is proud of) you would never know he was born with a defect. He may be behind his peers physically, but he is just as smart as they are."
- "Let them know they are no different."
- "Very good quality. Besides his dairy allergy, he rarely gets sick. He's very active, but disciplined and very sweet and kind to others. Loves Jesus, his family, his friends. He's very creative and can build amazing things with legos or whatever's around."
- "Fantastic quality of life"
- "She has a normal day to day life to me. She is very energetic, outgoing, and beautiful. Her life is wonderful. She I hope will one day be a career woman and mother."
- "He is very healthy and happy. He hasn't had any problems related to the gastroschisis."
- "Good. He's a happy 5 year old child who like normal kids loves life. It really has not affected him at all."
- "The only problem my baby has is constipation. She really never complains of anything."

- "Now at 3 years old, he is a normal boy with a lot of energy and is like any other boy his age but he suffered a lot his first year, God bless him."
- "He is just like any other kids his age. Very outgoing kid."
- "healthy and happy"
- "very healthy and active"
- "My daughter has a wonderful quality of life with limitations. We don't allow her to participate in any activity where she can be injured severely in her abdomen."
- "My daughter has two cousins her age and she has done everything they have developmental at the same time as them or before them."
- "Great. Very few problems from birth to 2 years of age. At the age of 3 (now) she is learning, growing, and blessed as all other kids (I guess)."
- "Hard to handle at times. Scared he can hurt himself."
- "It's normal like nothing's wrong. He's special and we acknowledge that but we live life day to day."

3. What advice would you give to a parent whose child just received a diagnosis of gastroschisis?

- "Stay positive and obtain more info on what to do after being diagnosed. Do research as that too can be helpful depending on their case."
- "Don't be scared."

- "Love and care"
- "Let God handle it."
- "Learn all you can about it. Don't be afraid of pictures you may see on internet or in books. Don't make a big deal of it and don't overprotect."
- "Not to worry. Child will be as normal as any other kid and grow up to do everything. Child will not miss out on anything."
- "Just to be patient that everything is going to be okay. Just to love your child more."
- "Have patience and have a single pediatrician that is recommended by the hospital and that it takes time, but it's worthwhile because they are normal children."
- "Research for yourself. Look at the pictures, prepare yourself for the sight of your child. Be open and ask as many questions as needed to ease your mind. No worries. My son is perfectly fine. NO hindrances whatsoever from gastroschisis."
- "Not to be scared, be there for your baby every day, talk to them, read to them, and hold them as often as you can. Let them know you are there."
- "It's good if they put themselves in the hands of doctors that know what they are doing and let them save the life of their children."
- "It is a hard thing to go through the first 2 years, but then things get better and your child will live a normal life. The surgeon told me out of all the things for your child to have go wrong that this was the best thing to get. He was right!"

- "The road to recovery will be rough at first, but the long-term outcome and quality of life for a child born with gastroschisis is good. It's essential to have a good support team and talk with other moms that have had children with gastroschisis."
- "We prayed daily over our son. We listened to the doctors and followed what they said."
- "Do not panic! Success rate is good-after, life is usually better. Child will have some hiccups but will live a happy life."
- "Everything will be fine. Out of all the birth defects out there, this is the one to have if you had to pick one. It can easily be fixed and your child will live a normal life."
- "Get educated about what gastroschisis is and with patience and learning they will see their children move forward."
- "Be encouraged because your child can have a healthy and normal life."
- "Be strong and always keep head up. Always think positive."
- "That they don't worry so much. The medicine is very advanced and the doctors are very prepared."
- "Expect the best. Prepare for the worst and continue to pray. Faith and attitude make a world of a difference."
- "You will know your child and what s/he needs. Listen to your gut."
- "That everything is going to be okay and that they have a lot of faith that God is going to heal the child."

- "I would tell them that their road ahead will be bumpy and scary, but if they pay attention to their child, they will know exactly what he/she needs and their life can be happy. Different, but happy."
- "Don't be afraid. There are so many doctors now who know what to do. Have faith. God will take care of the rest. My boy was in NICU for 6.5 weeks with this. He is healthy, had no issues due to defect."
- "Patience, it's always distressing knowing that your child has a problem, but thanks to God, my son is okay."
- "It's something very painful to find out that your child has this problem, but nowadays science is very advanced and there are many possibilities for our children to have a good quality of life."
- "Get Dr. [B] to be your baby's surgeon. He is AWESOME. Even though it is a very shocking and traumatic result to have while pregnant, it is all fine once the baby is born and is operated. It is tough to have to wait until the baby is out of NICU, but in the long run, it is all for the better of both the child and the parents' life. Children with gastroschisis live a good lifestyle as any regular child. Keep the baby that it will all turn out good."
- "The specialist/surgeons were very thorough with explaining diagnosis and neonatal surgery. They keep a very close eye on the BD with weekly checkups."
- "Be prepared but know that as bad as it may seem, it will be okay. It's something minor in the big scheme of things."

- "Limit your own personal research. I frightened myself to death with too much information, spending hours on the internet. Tour the hospital neonatal unit. Ask questions about the tubes, monitors, IVs, etc. Most birth defects can't be repaired and then have no long-term effect. This one can, not always, but often."
- "Hang in there. It gets worse before the good comes. Always know in your head it can happen again. Watch his/her activities. Good luck."
- "Do as much research as desired on the diagnosis and don't hesitate to ask your specialist any questions. Also, make sure that your decisions on your hospital are adequate to take care of babies born with this problem. But, everything will be fine and your little miracle has been blessed with strong parents because God doesn't put everyone in these situations. You're NOT ALONE!!"
- "Each case is different, but with prenatal care watching careful and determining when to deliver helps make sure baby is not in stress. Find a hospital that has a good team of pediatric surgeons. Be patient each day in the first year, could be challenging."
- "Trust your doctors. Everything will be fine!"
- "Never be afraid to ask questions! The more you know about the defect and your child's prognosis, the easier things will be; therefore, you are in a better position to care for your child and be able to know if his/her gastroschisis is creating problems in the future."

- "Follow the doctor's plans. It's all in the baby's time. It may be a bumpy road, but hang in there. It will get easier and hang on to God. That's what kept us going."
- "Not to worry. Always cherish him/her. Never make them feel different from other children."
- "Doctors have to give you the worst case scenarios so don't stress right away. Have faith in your doctor and NICU nurses."
- "I did not know D had gastroschisis until he was delivered/born. His entire stomach, small, large intestines were on the outside. We feel strongly that my regular exercise and healthy diet were large contributors to him being released from the hospital after only 2 months. Dr.'s and RNs thought he would be there 6-9 months."
- "A child with gastroschisis is a child that God placed in your hands because you can handle it. Be ready for the good and the bad, the tough and the simple. Prepare for the worst while praying for the best. Children are a blessing."
- "Try not to stress about it. If possible, opt for primary closure."
- "Stay calm, breath. Everything is going to be ok. At the time you don't know what to do and it's ok to be scared, but stay strong for your child and when it's all done with you're going to see it wasn't half as bad as you thought it was going to be."
- "I would say "Don't worry. Everything will be ok. Just be prepared to see a doctor near you. If in Midland they'd send you to Dallas. But the doctors are great. Calm down and don't stress too much."

- "My first advice is put your child in God's hands and God is going to help you endure this and then trust in the doctors; they do a good job."
- "Listen to your OBGYN specialist; don't believe everything you read on the internet because every child is different."
- "Don't look on the internet."
- "Prepare yourself as much as you can. Don't be afraid to ask questions."
- "The world isn't over, with time and patience the emotional roller-coaster will bring you and your child together. Technology and doctors will do what they can and you can only hope for the best."
- "Your child can go on to function normally but there are certain activities that require you as a parent to watch or monitor more closely than other parents."
- "It's scary but your child can have a normal childhood and will have a special man-made belly button."
- "Very difficult to understand, but take it a day at a time and receive all knowledge available."
- "To watch every step he/she takes and watch them a lot more just to make sure he/she doesn't hurt themselves"
- "It's scary. Just do your best to hope that everything will be okay and don't stress."

4. Any other comments for the researchers looking into gastroschisis:

- "Request: if at all possible, could I receive feedback or guidance on what or where my son should be taken or seen based on my answers?"
- "I his mother was also born with severe gastroschisis. I developed intestinal malrotation and occasionally have severe scar pain. Other than that I'm good. They did not know my son had it until his birth. His appendix was removed and exploratory surgery performed."
- "Mother should never lift heavy objects, avoid caffeine, take care of yourself."
- "We were luckier than most."
- "The problem that we cannot fix: [she] is 12 and still wets the bed 2-3 times a week."
- "My son plays soccer. He is the goalie. Also, plays basketball and he is in band and a good swimmer. The only thing I suggest is to keep children in shape."
- "That they study what the cause is that causes this because there isn't much information for the people or for parents. What is the cause?"
- "Interested to know about damage to abdominal wall muscles. What happens to them? My son's seem to be misshapen around his scar. Doesn't seem to cause discomfort or lack of core strength, but I want to know are they different because of the initial surgery? I would love a follow-up if possible, as well as to participate in any other way to help with families of gastroschisis babies. Please see contact info if needed: [. . .]"

- "Please send all further questionnaires to: XXXXX. I might take a while to respond but I'm glad to help with this."
- "I know of two other cases where the child born with gastroschisis also developed strabismus. Possible link or just a side-effect of being born premature due to the gastroschisis?"
- "We were told that he had an omphalocele then saw the specialist and were told it was gastroschisis and completely fixable, not to worry. [He] had surgery 3 hours after birth and was out of the hospital in 10 days."
- "It would be wonderful if there were some guidelines set as far as the aftercare of children born with gastroschisis are concerned. Most pediatricians are not sure what to tell us. We tend to know more than they do. A lot of times we aren't sure if her tummy troubles are gastroschisis related or viral. Thank you for doing this research."
- "It seems these kids are very intelligent. Scar is getting bigger but he is proud. God gave me a miracle."
- "Nothing more than to thank them because thanks to all the studies they have done and put in practice, today I can have my daughter with me and enjoying a good quality of life. Thanks and God bless them."
- "As a teenager (prior to pregnancy), I experimented different types of recreational drugs and when I was pregnant, I was working as a dental assistant and was exposed

to x-rays radiation. I would like to one day find out if these facts have anything to do with a fetus developing gastroschisis."

- "Find it as early as my sonographer did 12 weeks. First sonogram to find out how far along I was was a vaginal sonogram."
- "I am convinced that taking over the counter sudafed early in pregnancy caused this and I have other gastroschisis mothers on the internet that say the same. Also seems to be very prevalent for very young mothers and I was 36 years old when I delivered."
- "How to improve surgery so it won't have to happen again?"
- "Words can't express how happy I was to do this survey. I think what y'all are doing is amazing. It's about time this medical condition is looked at closer, so expectant families feel supported, not lost, scared, and confused!"
- "We had the best case scenario except a staph infection after surgery."
- "Thank you! When I first received the diagnosis for my son at 16 wks gestation, I poured through literature and the internet looking for answers and information. Unfortunately, I didn't find too much. Thankfully, the pediatric surgeon and neonatologist at my hospital were very familiar with the defect. I pray your research yields more information and answers for future parents who find themselves in the same position where my husband and I were almost 5.5 years ago."
- "No one to this day told us what to expect after the hospital. D did return for a 2 week stay and they discovered the start of an infection in the lining of the intestine."

Apparently very common- No one told us! I know someone who is 23 years old who was born with gastroschisis. She had to have surgery at 21 years. I still don't know if we should be seeing a GI doctor from time to time for checkups. We worry about what, if anything, to expect in the future."

- "My child had a bowel obstruction in utero. Is there any way that can be prevented? Is there any way the wound could be closed neater? My daughter's scars are very big. Six weeks after leaving the hospital my daughter got RSV. She was very small, skinny. A colonoscopy was done at the hospital because she was so small. At that time she did have ulcerative colitis. The kidney specialist was for the acid levels in her blood were too high. The first two years of her life were the most difficult time of her gastroschisis. Thank you so much for caring about these children."
- "I'm not sure if that has anything to do with why she got gastroschisis but I was always stressing, crying, and worrying. But mainly crying, depressed, and not eating right because her dad was emotionally abusive plus other things but never physically abused."
- "I'm happy that they are concerning themselves with this because it's difficult for one as the parent to ask for advice and they don't have an answer. Thank you for caring and God bless them."
- "No comments. Keep up the good work. Thanks for all that y'all do."
- "Each patient is different, but as I see in my child she has excelled at everything she is involved in."

- "More specific questions will better help understand. By the questions you asked, you wouldn't really understand my child's true journey of her gastroschisis. Much more to the story."
- "I would like to know if the average life span of a child born with gastroschisis is different than a normal child born without gastroschisis."

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Vita:

Roya Michele Mostafavi was born in Memphis, TN on December 12, 1987 to Ali and Nasrin Mostafavi. She has one older sister, Mariam. Roya grew up in Memphis and, after completing her education at Briarcrest Christian High School, she attended the University of Memphis. She graduated in 2009 with her Bachelor of Science in Biology. In August 2010, Roya entered the Genetic Counseling Masters Program at the University of Texas Health Science Center in Houston. She will graduate in May 2012.